

PEDIATRICS

Handwritten Note

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Name: _____

Subject: _____ **PEDIATRICS**



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PEDIATRICS

Neonate - First 4 wk. of life.

↳ Early - 7 days, cause of death - Prematurity

· Late - 7-28 days, - Sepsis.

Term - 37-42 wk gestation

Preterm - < 37 wk

Post term - > 42 wk

Normal wt \rightarrow 2.5-4 kg

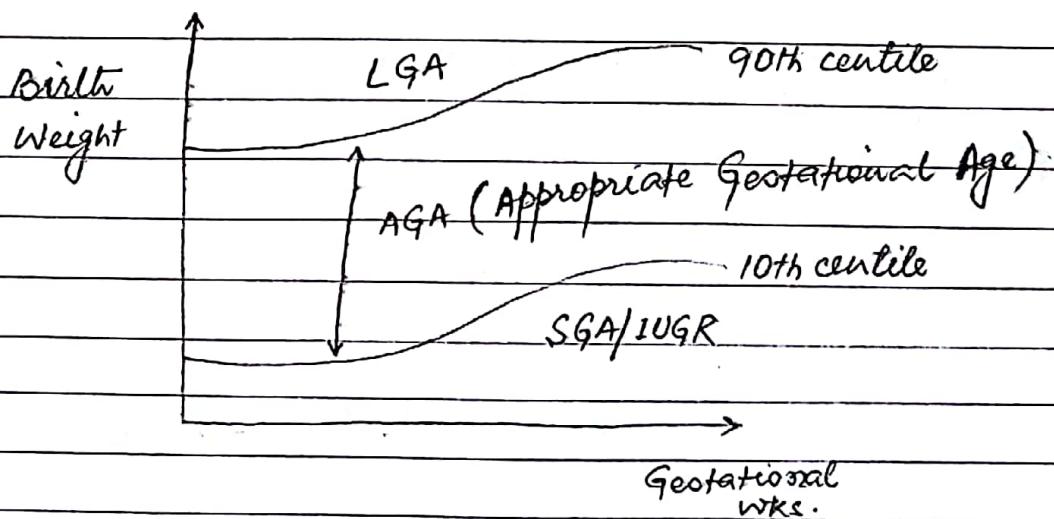
LBW < 2.5 kg

VLBW < 1.5 kg

ELBW < 1 kg

Macrosomia > 4 kg.

Lubchencho chart:



Constitutional:

- LGA (Large for Gestational Age)
- Short stature.
- Delayed puberty.

- All term baby has 6 fontanelle:

- Anterior \rightarrow ~~closed~~ ~~as~~
- Posterior
- 2 sphenoid
- 2 Mastoid.

Posterior fontanel - closes at birth.

open in 3% baby.

Anterior fontanel - diamond shaped.

2.5 x 2.5 cms.

At level

Pulsatile

closes at 18-24 months of life.

Craniosynostosis:

all

Early closure of fontanel.

Complication:

- Microcephaly
- IGT \uparrow \rightarrow optic atrophy.
- cosmetic.

Rx - Cranieotomy

Syndromes associated - Apert

Crouzon] AD

Pfeiffer

Carpenter - AR

Delayed closure of fontanel:

- Rickets
- Down's
- Hypothyroidism
- Hypophosphatasia
- cleidocranial dysostosis.

Q. 2 wks baby, Hypotonia, hypothermia, umbilical hernia, constipation, physiological jaundice prolonged.
 ↳ Congenital hypothyroidism
 (humeral head - Epiphyseal dysgenesis)
 (46④ epiphysis of bone)

Neonatal Screening:

- TSH; T_4
 - Delayed rise of TSH
 - TBG deficiency (TSH \uparrow ; $T_4 \downarrow$)
 - Best time: after 48 hrs (72 hrs)

M/c/c of congenital hypothyroidism - 85% agensis/ dysgenesis of thyroid gland.

Congenital hypothyroidism:

Prevalance = 1: 2000

Girls > Boys.

Neonatal screening : Continue

TSH; T_4

- Cold temp^r
- Physiological TSH surge 48 hrs.
- APP $>$ 48 hrs to 6 days.
- In OP Ghai \rightarrow 3-5 days



\rightarrow Sample obtained \rightarrow Heel prick.
 Safe area \rightarrow Side prick.

- pt in shock, venous access can't get in 60 sec.
- M/c, ^{other} site - Near tibial tuberosity.
 Upper end of fibia
 Lower end of femur.
- In shock \rightarrow i.v. fluids \Rightarrow 20 ml/kg bolus NS.

Most easily assessible venous route - Umbilical Vein.

Phenylketonuria: Deficiency of Phenylalanine hydroxylase.

Phenylalanine

PKU \ominus ~~PKU~~ Phenylalanine hydroxylase.

~~PKU~~

Tyrosine

\downarrow Tyrosinase

DOPA

\downarrow

Dopa quinone

\downarrow

Melanin

\uparrow Phenylalanine \rightarrow Toxic to Brain \rightarrow so in PKU High Phenylalanine

- Child Mental Retardation

20 mg/dl

- Developmental delay.

- Exaggerated Reflexes

- Microcephaly.

Rx - PKU

- Supplement Tyrosine
- Restrict Phenylalanine
- Now Tyrosine essential.
- Aim: Serum Phenylalanine $< 6 \text{ mg/dl}$.
- Lifelong.

Q. About PKU Rx, first step -

A) Stop the substrate of the enzyme

B) Supplement the enzyme.

C) Reduce the substrate of the enzyme

D) Provide deficient protein.

Maple Syrup Urine disease (MSUD):

- Deficiency of α -keto acid branched chain dehydrogenase.
- High Valine : Leucine : Isoleucine Blood & CSF.
- Coma
- Severe acidosis.
- We do dialysis - To remove Valine : Lev : Isoleucine.

Neonatal Screening : Consilice

- Tandem mass
 - Spectrophotometry \rightarrow metabolic
- Cystic fibrosis
- Congenital adrenal hyperplasia
- G6PD deficiency.
- Biotin deficiency.

ANTHROPOMETRY : TERM BABY

Length - 50 cms

Head circumference - 35 cm

$HC > CC$ but not more than 3 cm.

- $HC > CC$ (> 3 cm) \rightarrow Congenital hydrocephalus.
- \rightarrow Asymmetric IUGR baby.

$CC = HC \rightarrow$ at 9-12 months

By 1 yr ; $CC >> HC$

Upper segment : Lower segment (US : LS)

New born = 1.7 : 1

At 10 yrs = 1 : 1

Adult = 0.9 : 1

Achondroplasia : Short limb dwarf (US/LS \uparrow)

Hypothyroidism : US/LS \uparrow (disproportionate short stature)

Meconium :

95% pass meconium at 24 hrs.

99% " " " 48 hrs.

Cause of delayed passage meconium

- Imperforate anus

- Hirschsprung / aganglionic

↳ Rectum biopsy.

- Meconium ileus → Cystic fibrosis.

↳ Small intestinal obstruction.

Q 48 hrs baby has not passed meconium. Next Ix

A) CFTR gene test

B) Sweat chloride

C) Manometry

✓ D) Lower GI contrast study.

↳ △ Hirschsprung

Treat meconium ileus.

Delayed Urine :

- B/L Renal agenesis

↳ Maternal oligohydramnios.

also cause ↓

Post. Urethral valve

B/L pulm. hypoplasia

↓

POTTER'S Sequence.

Potter's face - Nose pinched in

Retrognathia

Micrognathia

Bag & mask → Pneumothorax.

Q If a baby has not passed urine in 1st 48 hrs, Next Ix
 → USG

Q 3 days old, c/o - Weak dribbling urine stream.

O/E - palpable distended bladder.

Δ → Posterior Urethral valve.

IOC : MCU (Micturating Cysto Urethrogram).

Rx : Cystoscopic Fulguration

SGA / IUGR :

Complication : TORCH infection / Chromosomal disorders.

Symptom - Asphyxia → causes MAS

↓ (Meconium Aspirated Syndrome)

PPHN (Persistent pulmonary HTN of Newborn).

Severe IUGR - Pulmonary hemorrhage

Limited stores - Hypoglycemia, HypoCa; HypoMg.

- Polycythemia

↳ bcoz in IUGR erythropoietin is very sensitive to hypoxia.

- Neutropenia

- Thrombocytopenia.

Q Full term small for date babies are more disposed to -

a) HyperCa

b) CNS infection

c) PDA

common in preterm.

✓ Hypoglycemia

Symmetric IUGR

Asymmetric IUGR

Cause	Chromosomal/ Torch	Maternal Complications
Cell no.	↓	↑
Cell size	↓	↓
Brain	↓	↑ N spared. HC > CC > 3cm Brain/liver ↑

PONDERAL'S INDEX:

$$\frac{wt (gm)}{Length (cm)^3} \times 100$$

< 2 → Asymmetric IUGR

≥ 2 → AGA/ Symmetric IUGR

Normal Neonatal phenomena:

Milia: Distended Sebaceous gland on face & nose.

Erythema Tonicum: Erythema on face & trunk
2-3 days of life.

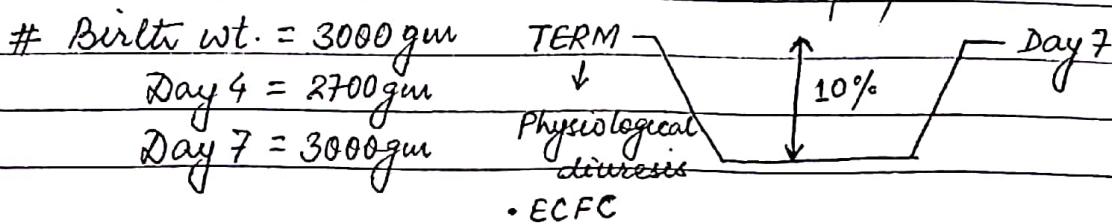
Stork bites: Pinkish gray capillary hemangioma
on back & buttocks.

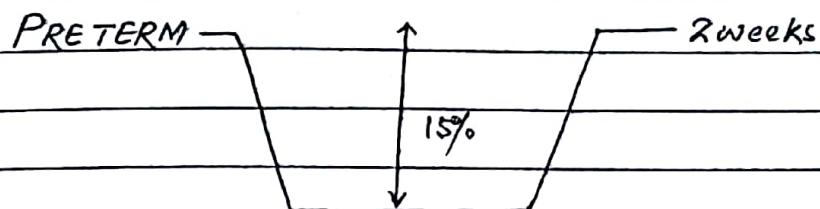
Epstein Pearl: Epithelial inclusion cyst on palate
& prepuce.

Natal teeth (Pre deciduous teeth): lower incisor position.

~~With~~ Withdraw vaginal bleeding: On 5th-7th day.

Acrocyanosis (Peripheral Cyanosis): Limbs cyanosed
Lips pink.





IUGR \rightarrow do not lose wt (ECF compact)
 \hookrightarrow wt. stable for 1-2 days then wt. gain.

Q. Not normal in a newborn?

- A) Proteinuria \rightarrow in ELBW
- B) Glucosuria
- C) 1-2 pus cells/hpf
- ~~D) Bacteriuria~~

Neonatal Reflexes:

① Moro Reflex:

1st phase - Abduction of shoulder joint
 Extension of elbow joint.
 opening of fingers

2nd phase - Adduction & flexion.

- Appears 28-32 wks gestation.

• Adduction / complete \rightarrow 36-38 wks.

- Disappears at around 2-3 months of life.

- Persistence beyond 6 months abnormal.



Cerebral palsy.

$\frac{1}{2}$ - Asymmetric moro's

- Brachial plexus injury.
- # clavicle
- # Humerus
- Hemiplegia.

Early hand preference is always abnormal

- Hemiplegia at other side.

- 95% cases → Hemiplegia at Rt. side.

Exaggerated Moro → HIE-1

ATNR (Asymmetric Tonic Neck Reflex):

Side of face - Extended

Side of occiput - Flexed.

Onset - 35 wks

Fully developed - 1 month

Duration - 6-7 months

Do not ~~roll~~ roll (Rolling start when this reflex disappears)

Disappears - 6 months

STNR (Symmetric tonic Neck reflex):

Neck extended

→ Zone ↑ UL

↓ LL

Neck flexed

↓

Zone ↓ in all limbs.

- Not present at birth.

- Appears 4-6 months of life.

- Disappears 8-12 months of life.

- Child start to crawl when this reflex disappears.

Parachute reflex:

- Not at birth

- Appears 6-7 months of life.

- Well developed at 10-11 months of life.
- Persists life long.

LANDAU Reflex:

- Appears at 3 months of life
- Disappears at 1 year of life.

On ventral suspension - spine strengthens / straighten.

Child get out of flexion attitude by this reflex.

GRASP Reflex:

- Appears 28 wks of gestation
- Well developed 32 wks
- Disappears 3 months of life.

Sucking & Rooting reflex:

< 28 wks absent.

At 28 wks - Some sucking bursts.

32 wks - Appear

34 wks - Co-ordination

Q

31 wks ; 1500gms ; Feed - ?

A) Enteral → NG tube (Expressed Breast Milk)

B) Enteral + i.v. fluids

C) IV fluids.

D) TPN.

Q

33 wks ; 1500gms ; Feed - ?

↳ Enteral → Katori / Paladay / Spoon

(Expressed Breast Milk)



AIIMS NICU Protocol 2014 :

> 34 wks - Breast feed

32-34 wks - Katori (Expressed breast milk)

< 32 wks - NG tube (" " " ")

< 1200gms - i.v. dextrose + minimal enteral feeds 10-15 ml/kg/day.

Rapid enteral feed can cause - Necrotising Enterocolitis.

Fetal alcohol Syndrome :

- Skin folds at the corner of the eyes
- Low nasal bridge.
- Short nose
- Indistinct philtrum (groove b/w nose & upper lip)
- Small head circumference (microcephaly)
- Small eye opening!
- Flat mid face / midfacial or maxillary hypoplasia.
- Thin upper lip.
- Septal defects < ASD
VSD.

Q Ass. C ~~and~~ fetal alcohol Syndrome except -

A) Microcephaly

B) Overgrowth

C) Flat face

D) Small palpebral fissure.

PREMATURITY

Respiratory System: RDS (Respiratory distress Syndrome).

Chronic lung disease /

Bronchopulmonary dysplasia

O₂ dependence on 4 wk of life

Rx: Home O₂ therapy.

Prevention: Nasal CPAP.

Vit. A supplementation.

Furosemide

Keep underhydrated in ICU.

CNS: ① Apnea (> 20 sec); or any period of ass. c. central cyanosis & bradycardia.

Q M/c signs of acute hypoxia in neonates

A) Bradycardia

B) Tachycardia.

Types of Apnea: 3 types

① Central: asphyxia; preterm.

② Obstructive:

all newborn are obligate nose breathers till 4 months of life.

Q: Full term newborn

episodes of cyanosis - worsen when feed.

Seems better when crying.

↳ Chonchal atresia → B/L, posterior.

③ Mixed (M/c)



Apnea of prematurity:

Risk: < 28 weeks $\rightarrow 100\%$

Onset: 1-2 days; never > 7 days.

Rx:

1st step: Nasal CPAP

Methylxanthines - Aminophylline (^{Narrow therapeutic range})
Caffeine Cilrāte (DOC)
↳ wide margin of safety

L Loading dose of Aminophylline: 5-6 mg/kg.
followed by maintenance dose
1-2 mg/kg every 6-8 hrs.

Caffeine Cilrāte: Loading dose \rightarrow 20 mg/kg

Maintenance dose \rightarrow 1-5 mg/kg/day

② Intracranial hemorrhage

(Only subdural hemorrhage is common in term baby).

- Capillaries in the subependymal germinal matrix is fragile; so they rupture.

Preterm; sudden pale; Shock, fontanel bulging; seizures - Intra ventricular hemorrhage (IVH)



Risk IVH preterm - < 1500 gm $\rightarrow 30\%$

[50% IVH \in 24 hrs.

[75% " " 72 hrs.

IOC for newborn having seizure \rightarrow Trans fontanel USG

Term; Breech \rightarrow IVH

Instrumental delivery \rightarrow IVH

Prevention of IVH:

- Antenatal steroids
- Low dose indomethacin to baby.
- Prevent acidosis, infections in baby.

③ ~~Asphyxia~~ Asphyxia

Preterm

Term

Periventricular
leucomalacia (PVL)

Parasagittal injury

Power: UL > LL

Spastic quadriplegic cerebral
palsy.

Diplegia
(upper limbs strong
LL are weak)

MRI: Coronal Section
- parasagittal injury.

↳ Spastic diplegia type
of Cerebral palsy.

• Mentally Retarded

Q. M/c sequel of Periventricular
leucomalacia in preterm
- Spastic diplegia

MRI: PVL (IOC)

- less white matter
- Shrinkage Ventricle.
- Mentally Retarded.

Stalis marmoratus: diffuse neuronal loss following asphyxia.

APGAR Score : 1, 5, 10 minutes

- doesn't help in resuscitation.
- 5'; 7'; 10' low → Worse neuronal outcome.

0 1 ; 2

A = Appearance Central cyanosis/ Acrocyanosis Pink.
Pale

$P = \text{Pulse (HR)}$ 0 <100 >100

G=Grinace *No* *Grinace* *Crying*

A = Attitude Extended Mid Flexed

$R = \text{Resp}^r$ effort Apnea Gasping Crying.

NORMAL - 7-10

Moderate to severe asphyxia; out of hospital CPR

ischemia reperfusion injury

Free radical damage.

Rx - Therapeutic hypothermia/ Selective head cooling:
 33.5°C \in in 6 hrs of life; Keep for 72 hrs.

preventive

Hypoxic Ischemic Encephalopathy (HIE):

Injury to brain at severe asphyxia.

Sigors	STAGE 1	STAGE 2	STAGE 3
• Loss of consciousness	<u>Hyperalert</u>	<u>Lethargic</u>	<u>Stuporous; Coma</u>
• Muscle tone	Normal	Hypotonic	<u>Flaccid</u>
• Posture	Normal	Flexion	<u>Decerebrate</u>
• Tendon reflexes/ clonus	<u>Hyperactive</u>	<u>Hyperactive</u>	Absent
• Myoclonus	+nt	+nt	-nt
• Moro reflex	<u>Strong</u>	<u>Weak</u>	-nt
• Pupils	<u>Mydriasis</u>	<u>Miosis</u>	<u>Unequal, poor light reflex.</u>
• Seizures	<u>None</u>	<u>Common</u>	<u>Decerebration</u>
• EEG finding	<u>Normal</u>	<u>low voltage changing to seizure activity.</u>	<u>Burst suppression to isoelectric</u>
• Duration	< 24 hr if progresses; otherwise may remain	24 hrs - 14 days	Days to weeks
• Outcome	Good.	Variable	Death, Severe deficit

DOC: Seizures in Newborn: Phenobarbital.

(Bolus, 20 mg/kg)



③ CVS: Hypotension; PDA (Patent ductus arteriosus).

PDA: Preterm \Rightarrow Asphyxia \rightarrow PGs

Term \Rightarrow Rubella infection \rightarrow Vessel wall defect.

\downarrow
Rx: Surgery

In preterm: Rx: NSAIDs

(Ibuprofen $>$ Indomethacin)

\downarrow
Less nephrotoxic

If Medical management fails \rightarrow Sx

C/F of PDA:

- 6-10 wks of life CHF
- Preterm baby, failure to wean off ventilator (hypoxia; CO₂ retention)

O/E: • Bounding pulses \bar{c} wide pulse pressure.

• Continuous machinery murmur at the upper left sternal border.

④ GI System: Necrotising Enterocolitis

⑤ Eye: ROP/ Retrolental fibroplasia.

⑥ Hypothermia; ^{Hypo} Glycemia; HypoCa; HypoMg.

⑦ Anemia; jaundice; Infection.

Necrotizing Enterocolitis (NEC) :

R/F: ① Immature Gut.

NEC: Susceptibility of premature infants

- Reduced proteolytic enzymes
- ↑ Gastric pH
- ↓ peristalsis
- ↓ motility
- Altered epithelial membr & tight junction
- Altered bacterial flora.
- ↓ mucous coat
- Altered mucous protein
- ↑ epithelial permeability.

- Sepsis toxins
- Top fed (cow milk)

② Mature cocaine

③ PPIs ; Anti H₂

④ Rapid advancement of feed.

Prevention: ① Antenatal steroids

- ↓ IVH
- ↓ PVL
- ↓ NEC
- ↓ RDS
- ↓ Neonatal mortality.

HUMAN MILK: ↑ proteolytic enzyme

↓ Gastric pH

↑ peristalsis

↑ motility

less pathogenic bacterial flora



- Altered mucus coat (Improves)
- ↓ epithelial permeability.

② Trophic feeds - expressed milk
10-15 ml/kg/day.

- ③ Avoid PPIs; anti H₂
- ④ Avoid rapidly feed advance.
- ⑤ Probiotics.

Q. 1 yr old infant; 10-12 episodes of watery stools/day for last 9 days. Along w/ Zn, which else should be advised -

- A) ORS & antibiotics
- B) ORS orally.
- C) ORS & low lactose diet
- D) ORS & low lactose diets & probiotics.

PROBIOTICS:

Prevents NEC - *Lactobacillus acidophilus* & *Bifidobacterium infantis*
to VLBW

- may use in Rotavirus:

Lactobacillus rhamnosus
& *Saccharomyces boulardii*

Modified Bell's Staging for NEC:

Stage Ia:	Suspected NEC	Distension; ileus; occult blood in stool
I _b :	Suspected	Gross blood loss.
IIa:	Definite	Focal pneumonitis
II _b :	Definite	HMT; Diffuse pneumonitis; Portal Venous Gas.
IIIa:	Advanced	DIC; Shock; Peritonitis
III _b :	Advanced-perforation	Pneumoperitoneum

Q. Neonate; distended abdomen & B/L gas shadow under the diaphragm.

Δ - NEC

↳ Ia 90% preterm

2nd - 3rd wk of life.

Q. Features of NEC are all except

A) Abd. distention

B) ↑ Bowel sound

C) Pneumoperitoneum

D) Metabolic acidosis

Rx: NEC

- Stop all oral feeds
- TPN (Glucose, AA's, Lipids)
- Antibiotics (Cefotaxime; Vancomycin; Metronidazole)
- Stage III - may require Sx.



Q. Child with NEC with perforation & poor general condition is treated with:

- A) Conservative t/t only.
- B) Frank drain & glove
- C) Laparotomy & resection anastomosis
- D) Extracorporeal membr oxygenation.

Stage III
Stable - Laparotomy
Unstable - Peritoneal drain

NEONATAL SEPSIS:

= Symptoms + Bacteremia

EARLY

< 72 hrs

LATE

> 72 hrs.

R/F - Maternal Fever < 7 days

M/c/c - Nosocomial

Foul liquor

M/c/c world - Coagulase -ve staph

PPROM: Chorioamnionitis.

In India - Klebsiella, S. aureus.

M/c/c world: Group B streptococcus, E. coli
(M/c)

Meningitis \Rightarrow CSF exam

Rx - Ampicillin + Gentamycin

- Cefotaxime + Amikacin

M/c/c India: Klebsiella, S. aureus

Sepsis screen for early diagnosis:

- ① TLC < 5000/cumm or > 20000
- ② ANC < 1500/cumm
- ③ PS for band cells / Immature neutrophile > 20% (I/T > 0.20)
& toxic granules
- ④ Micro ESR (8mm^(N) - 3 days of life) > 15mm fall in 1st hour
- ⑤ CRP: Procalcitonin +ve
- ⑥ Lumbar puncture (In late sepsis)



(7) Chest X-Ray

ANC = Neutrophil + Band cells

Q Lab finding in Neonatal Sepsis except -

- A) ↑ CRP
- B) Leucocytosis
- C) ↓ ESR
- D) Toxic granulated multilobulated nuclei

Duration of Antibiotics in Neonatal Sepsis:

Bacteremia = 10-14 days

Meningitis = 21 days

Arthritis, Osseous myelitis = 4-6 wks

Temp regulation of newborn:

Non-shivering Thermogenesis -

Brown fat

- Nape of Neck
- Interscapular
- Around kidneys & adrenal
- Around blood vessels (around mesentery).

Axillary temp:

- Normal $36.5 - 37.5^{\circ}\text{C}$
- Cold stress / mild hypothermia ($36-36.4^{\circ}\text{C}$)
- Moderate hypothermia ($32-35.9^{\circ}\text{C}$)
- Severe ($< 32^{\circ}\text{C}$)
- Hyperthermia $> 37.5^{\circ}\text{C}$ - mostly exogenous.

Prevention of hypothermia: KMC (Kangaroo mother care).



Q. All components are of KMC except:

- A) Kangaroo position
- B) Kangaroo nutrition
- C) Early discharge & follow up.
- D) Supplementary nutrition.

Q. When to stop KMC:

- When a child reaches 2500 gm wt.

Q. Delivery room temp - $> 72^{\circ}\text{F}$ (AAP)

$> 25^{\circ}\text{C}$ (WHO)

$\hookrightarrow (25-28^{\circ}\text{C})$

ALIMS NICU protocols

- $< 28\text{ weeks}$ / $< 1000\text{ gms}$ \rightarrow put baby in Polythene bag.

reduces convection

- Incubator \rightarrow Reduces convection.

Q. Mechanism of heat transfer in overhead radiant warmers :-

Radiation, + ↓ convection loss.

Hypothermia:

CNS: Asphyxia

CVS: Asphyxia Cardiomyopathy

Lungs: ARDS

Kidneys: Asphyxia ATN

Hypoglycemia; HypoCa; HypoMg.

Retrobulbar Fibroplasia /

ROP (Retina of Prematurity):

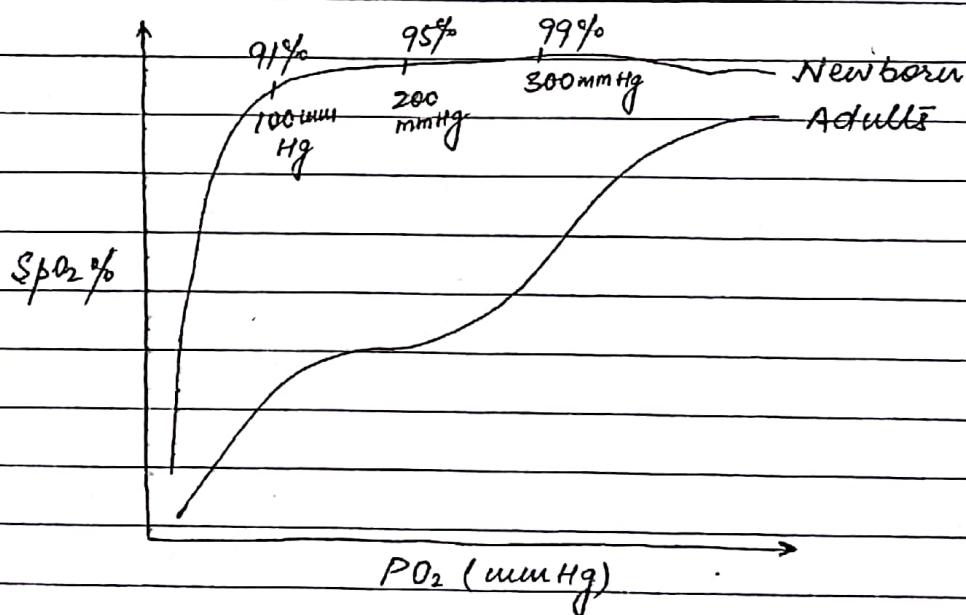
R/F: Preterm.

Higher flow O₂.

- Proliferation, dilatation & tortuous vessels.
- Tractional retinal detachment.
- ROP stage I-V: 'plus' - Blindness.

Stage IV: Incomplete tractional RD

Stage V: Complete " "



WHO targets SpO₂ = 91-95% in preterm.

Inv. of ROP

BLINDNESS: Regular indirect ophthalmoscopy.

AAP ROP guidelines - (Risk < 30 wks / < 1500gms)

Gestation (wks) 1st visit to ophthalmologist

Post menstrual (wks) Interval (wks)

22

31

9

23

31

8

24

31

7

25	31	6
26	31	5
27	31	4
28	32	4
29	33	4
30	34	4

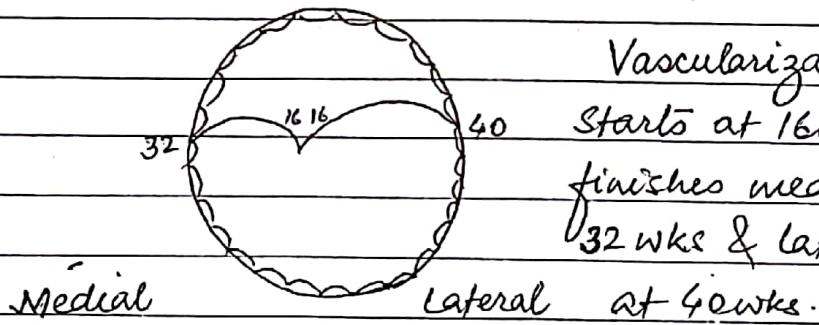
Q. Pediatrician in a district hospital calls ophthalmologist for

a) Newborn c Respiratory distress

~~b)~~ Newborn 28 wks gestation

↳ After first visit, baby has to go to ophthalmologist, every 2 wks till his/her eye look like term retina.

Retinal Vascularization: Left eye.



Vascularization of Retina
Starts at 16 wks,
finishes medially at
32 wks & laterally
at 40 wks.

Rx: ROP

- Laser Photo coagulation/ Peripheral ablation
- Type I ROP (all plus disease)
- Cryotherapy.
- Stage V → Retinal reattachment.

New drug - Bevacizumab → Anti-VEGF; resistant.

Respiration:

Tachypnea: RR > 60/min.

Silverman Anderson Retraction Score:

Feature	Score 0	1	2
Chest movement	Equal	Respiratory lag (upper chest inspiration)	Paradoxical/ Saw Respiration
Intercostal Retraction	None	Minimal	Marked
Xiphoid Retraction	None	"	"
Nasal Flaring	None	"	"
Expiratory Grunt.	None	Audible c Stethoscope	Audible

Downes scoring for Respiratory distress:

Feature	Score 0	1	2
RR (per min)	< 60	60-80	> 80/ apneic episode
Cyanosis (central)	None	In room air	in 40% O ₂
Retraction	"	Mild	Moderate - Severe
Grunting	"	Audible c Stethoscope	Audible c Stethoscope
Air entry (mid-axillary line)	Clear	↓ (Delayed)	Barely audible



M/c/c of RD → Surfactant deficiency

Surfactant → Phosphatidyl choline (65%) (Most imp.)

Phosphatidyl glycerol

Phosphatidyl inositol

Phosphatidyl ethanolamine

SP-A, SP-B, SP-C, SP-D.

Other protein.

↳ Homogenates - 20 wks gestation

Amniotic fluid - 28 wks gestation

Mature levels - 95% 35 wks.

RDS risk OC Degree of prematurity.

Risk of RDS:

< 28 wks - 60-80%

> 37 wks - < 5%

Infant of diabetic mothers, ^{at} term

Rare in IUGRs → Stress → Cortisol

Lechithin: Sphingomyelin ratio

> 2 → lung mature.

> 3.5 infants of DM mothers.

CXR: RDS (lungs) : Ground Glass appearance.
(white out lung).

Air bronchogram - classic feature of RDS.

(N) CXR seen in Early RDS.

$\text{Rx}:$ • Humidified O_2 (40-60%)
 - We don't give 100% O_2 bcoz of ROP.

• Nasal CPAP: mild - moderate distress
early in ELBW's

FiO_2 : Start \approx 40-60% \rightarrow room air

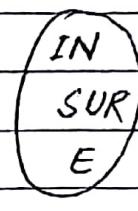
PEEP: 5 cm H_2O

• Intratracheal surfactant - Severe; as rescue.

- Intubate the baby

Give Surfactant

Extubate the baby



IN
SUR
E

Survanta - Bovine

Curosurf - Porcine

Infasurf - Calf.

Synthetic also available

Q

32wks, Preterm baby in emergency C.S.

Grunting, RR = 70/min. Best management of choice

A) Humidified O_2 by hood

B) Mechanical ventilation.

C) CPAP

✓ Surfactant therapy & mechanical ventilation.

Q

All occurs in RDS except:

A) Cyanosis

B) Occurs in preterm

C) More in IDM

✓ Treated by 100% O_2



Q. All true about CPAP except :

- A) Initiated $FiO_2 0.40 - 0.60$
- B) Used in apnea of prematurity.
- C) Improves compliance.
- D) Volume ; $FRC \uparrow [RDS FRC < CV]$
- E) Used prophylactically in ELBW's.
- F) All true.

Q. Term female, Birth wt. = 3.5kg, uncomplicated delivery.

Respiratory distress after birth.

CxR - Ground glass appearance.

On ventilation & given surfactant.

But cond'n deteriorates & hypoxemia increases.

H/o sibling dying in one week & similar complain. ECG & blood culture - N.

↳ A = Neonatal Pulm. Alveolar proteinosis

↳ Autosomal Recessive

↳ Mutation in Protein B (Rarely in c)

Rx : Early lung transplantation

Postmortem Rx - Pink eosinophilic material in lung.

- Idiopathic : 90%

Adult or acquired

IgG antibodies to GM-CSF

- Secondary - 5-10%

Hematological malignancy

Inhalational lungds.

Silicon

Titanium Oxide.

- Congenital : 2%

CT scan - crazy paving pattern
(Prominent intraalveolar septae).

Rx - Early lung transplant in neonates.

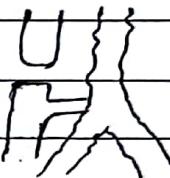
Adults → Broncho alveolar lavage.

Respiratory distress : Newborn

Tracheo-esophageal fistula (TEF)

M/c type - Type C

Distal TEF



Esophageal atresia

- Cyanotic newborn & frothing
- Aspirates gastric juice lead to pneumonia.
- Not a surgical emergency.

Rx - Keep him propped up.

- Suction catheter in upper blind esophageal pouch.

Diaphragmatic hernia:

85% cases - left.

pushed heart & trachea - to opposite side.

- Scaphoid Ab domen
- Barrel chest
- Mediastinal shift to right.
- Apparent dextrocardia
- Peristalsis on left chest.

Diaphragm - develops from septum transversum
& pleuroperitoneal canals.

↳ fail to close on left side

(Bochdalek hernia)

- < 5% are B/L



Q. Cause of death in Congenital Diaphragmatic Hernia:

A) Septicemia

B) Pulmonary hypoplasia - Left

C) Hemorrhage.

Bag & mask is absolutely c/i bcoz it will cause further abdominal distension.

↓
So elective intubation is done.

Baby born → Bag & mask & 100% O₂

↓

diaphragmatic hernia

↓

Intubated

Next step - NG tube to decompress the gut.

Born baby → Diaphragmatic hernia

↓

Intubated

↓

Heart further to right

Next step - Remove the tube
& Reintubate.

Q. Most imp. Prognostic factor in Congenital diaphragmatic hernia:

A) Pulm HTN - Persistent (PPHN)

B) Age

C) Time of Sx

D) Size of defect.

Prevention: PPHN

- Elective intubation
- $HFOV > 300-600/\text{min}$

Rx: PPHN

- iNO \rightarrow Pulm. vasodilator.
- Sildenafil PDE-5 inhibitor

New drug: Bosentan, Ambrisentan — Endothelin antagonist.

- $PGI_2 \Rightarrow$ Iloprost
- Amlodipine; ECMO (Extracorporeal membr oxygenation)

Transient Tachypnoea of Newborn (TTNB):

R/F:

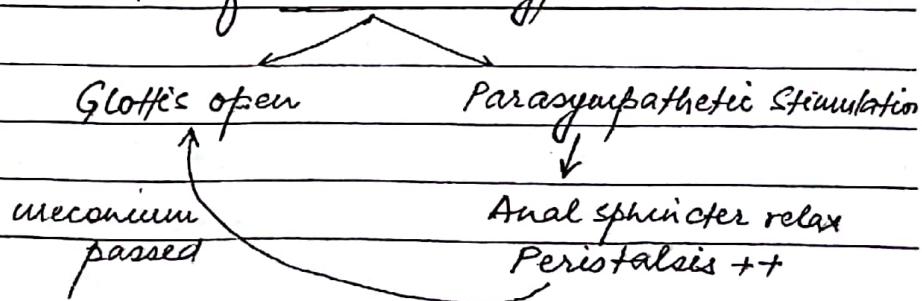
- Term by C.S. (lungs are wet) \ominus
- Macrosomia \ominus
- Excessive maternal sedation.
- Precipitous labour.

CxR — a prominent horizontal fissure
 ↳ Most specific feature.

- Benign cond'n
- Self limited: 48-72 hrs.
- FiO_2 requirement < 0.40
- Never require mechanical ventilator.

Meconium Stained Liquor:

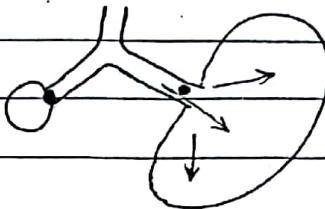
Meconium is marker of Perinatal Hypoxia



Perinatal hypoxia: Common ein posterum
scuz of VPI.

Meconium:

Physical -



- ball valve mechanism
- air leak 20-30%

Chemical - Irritant \rightarrow pneumonia

\hookrightarrow Impair surfactant funcⁿ.

Biological - Good culture media

Meconium Stained liquor.



Baby born

Vigorous

Tone is good

Resp effort is good

HR $> 100/\text{min}$

yes

No

Transfer

the baby to
mother

Catheter

in the nose



PPV \ominus 100% O₂.

Q

Sequence of Resuscitation

A) Mouth \rightarrow Nose

B) Nose \rightarrow Mouth

C) Mouth \rightarrow Nose \rightarrow Trachea

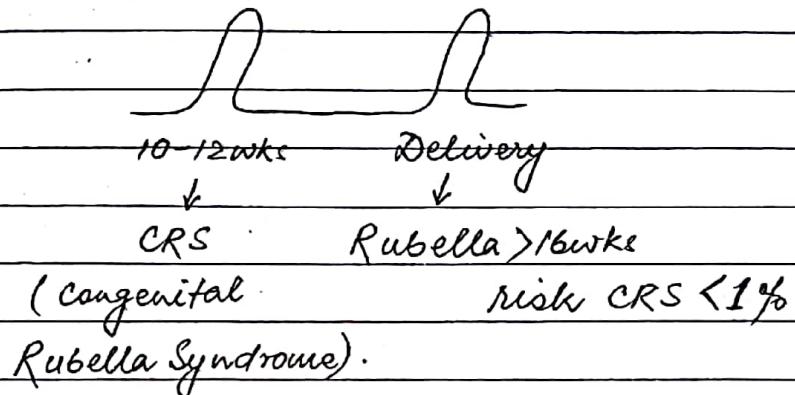
Intrauterine TORCH infections:

Others - HIV, HBV, Varicella, Syphilis.

Common features:

① - Asymptomatic

Rubella: Transmission - 2 peaks



② - If symptomatic

- SGA; failure to thrive
- Anaemia; thrombocytopenia
- Hepatosplenomegaly
- Unexplained Rash & cholestasis.

△ - IgM (evidence of infection)

IgG persisting beyond 6-9 months

HIV in infants

- Infants can't make IgM-HIV or IgA-HIV.
- Maternal IgG-HIV can persist in the baby for 18 months.
- HIV < 18 months: Diagnosis.

Best: DNA qPCR

P24 assay

Culture is difficult.

Intracranial calcification & Chorioretinitis] M/c/c - Toxoplasmosis - CMV

Toxoplasmosis

- 25-50%
- Choroid plexus calcification.
- Subependyma & Cadate nucleus calcification.
- ~~Hydrocephalus~~ Hydrocephalus
- Seizures.

CMV

- Periventricular Calcification.
- Atrophy of brain (cortical).
- Hydrocephalus ex vacuo.
- M/c/c of Non syndromic SNHL
- Seizures
- Microcephaly
- Mental Retardation.

Q. Pregnant lady; no complain. Mild cervical lymphadenopathy in 1st trimester. Prescribe

prevent vertical transmission of hydrocephalus & intracerebral calcification.
 (Spiramycin) but she was non compliant. Baby born

↳ Toxoplasmosis

DOC: Rx: Pyrimethamine + Sulfadiazine.

△: Best: IgM Immunosorbent assay.

Sensitivity of ELISA:

IgA > IgM

Q. True about transplacental CMV infection:

- It is M/c/c of non-syndromic SNHL.

Q. Doesn't establish Δ of CMV in neonate -

- A) Urine culture of CMV
- B) IgG CMV antibodies in blood
- C) Intra-nuclear inclusion bodies in hepatocytes (Owl-eye)
- D) CMV viral DNA in blood

by polymerase chain react.

Best specimen - Urine culture & Saliva.

CMV disease: Retinitis, colitis, pneumonia.

Rx : CMV

DOC : i.v. Ganciclovir (Severe, child, pregnant)

- Oral; Prodrug → Valganciclovir
- Resistant to oral Foscarnet.

Congenital Syphilis:

EARLY

- C in 1st 2 yrs of life.
- Mucocutaneous rash/rhininitis (Snuffles).
- Lymphadenopathy.
- Hematological (Autoimmune Anemia)

LATE

- After 1st 2 yrs.
- Hutchinson's Triad:
 - ① Hutchinson teeth/ mulberry molar - 1st lower molar.
 - Saddle nose, Frontal bossing, Olympian's brow.
 - Higoumenaki's Sign (sternooclavicular prominence)
 - Rhagades
 - ② Interstitial keratitis
 - ③ Nerve deafness. (SNHL)
- Cluttons joints (painless joints)
- ↳ Risk of injury.

Pseudoparalysis:

M/c/c - Scurvy

- Early syphilis
- Osteosyphilitic
- Septic arthritis.
- Hypokalemia → Hypotonia.

DOC - Penicillin G

→ 10-14 days



Rubella Syndrome :

Trisomy — Microcephaly (Mental Retardation)
PDA

Cataracts.

M/o eye manifestation of Rubella — Salt & pepper fundus

Cataract

Glaucoma

Micro-ophthalmia

HEART — PDA

Peripheral pulm. stenosis

VSD

ASD (Rare).

Q. Rubella embryopathy except.

- A) Deafness
- B) MR
- ~~C) AS~~
- D) PDA

Q. True about Rubella embryopathy except :

A) Diagnosed when IgM antibodies in child.

~~B) Infection after 16 wks results in major congenital anomalies.~~

C) Deafness, heart disease, Cataract.

Q. Hypoplastic limb — Varicella (Chicken pox) embryopathy during pregnancy.

Varicella Embryopathy -

- Skin Rash
- Optic nerve hypoplasia
- Brain - Cortical atrophy.
- LS plexus - Aplasia / hypoplasia limbs.

Mother get chick pox - 5 days before delivery
 or, \leq in 2 days of delivery.

↓
 Baby chicken pox illness.

Prevention: Varicella Zoster Ig to the baby.

↓
 even after 120 hrs of exposure.

Q Pregnant ; HBsAg +ve : no jaundice

$HBeAg$ +ve \Rightarrow 90% chance baby is carrier

& later in life Portal hypertension.

↓

Ascites, Splenomegaly, Varices

$HBeAg$ -ve \Rightarrow Anti $HBeAg$ +ve

then 10% chance of Vertical transmission

Give HB Ig baby to 12 hrs of life

HBV vaccine to baby \leq in 24 hrs of life.

Pricked by HBsAg +ve patient

Are you immunized?

~~Anti-HBc Ig~~ - Anti HBs Ab titre

Good $> 10 \text{ mIU}/\text{ml}$; High risk > 100

Incomplete vaccination / Not know titre



HBV + HB Ig

a/c to CDC guidelines.

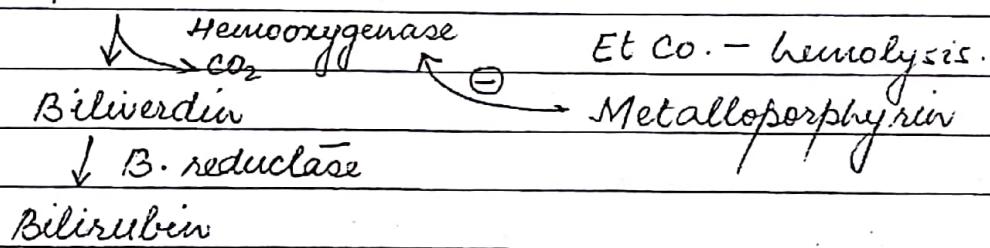
If titre is good - don't do anything.

CDC HBV DNA load > 1000 IU/ml

↳ Can't gain Surgical branch.

Neonatal jaundice

Mean



Bilirubin

1 gm of Hb = 34 mg of bilirubin

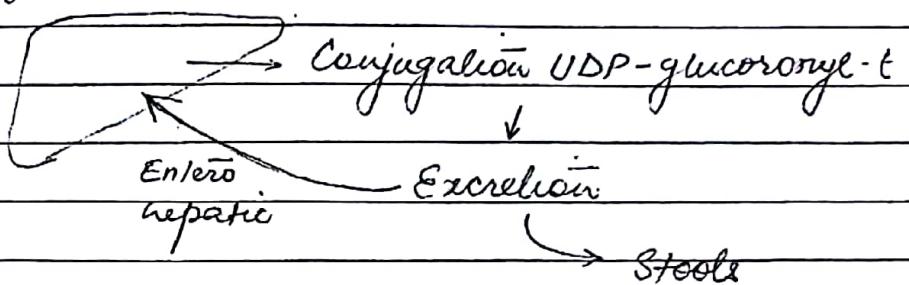
1 gm / dl Albumin binds to 8 mg bilirubin

- Unconjugate bilirubin passes BBB & cause jaundice & Kernicterus.
- In 1st 2wks, BBB is not developed properly.
- (N) S. Albumin = 3.5 - 5.5 g/dl
- Healthy term baby can bind 24 - 25 mg/dl bilirubin.
- Sick, preterm, risk factors can go into early Kernicterus.
- In adults ammonia crosses BBB in hepatic encephalopathy.
- Healthy Baby (< 1000 g) \rightarrow We start phototherapy ($5-7$ mg/dl Bilirubin)
 - ↳ Sick baby \rightarrow Bilirubin ($4-6$ mg/dl) - we start phototherapy.

Bilirubin



uptake by liver (γ -ligandin uptake)



$> 2-2.5 \text{ mg/dl}$ in adults \Rightarrow Yellow Sclera.

Kraunuer's zone:

progression of jaundice in new born is
cephalo-caudal
(Bilirubin)

Zone I (5 mg/dl) \rightarrow

Zone II (10 mg/dl) \rightarrow

Zone III (12 mg/dl) \rightarrow

Zone IV (15 mg/dl) \rightarrow

Zone V ($> 15 \text{ mg/dl}$) \rightarrow

↳ Danger Zone.

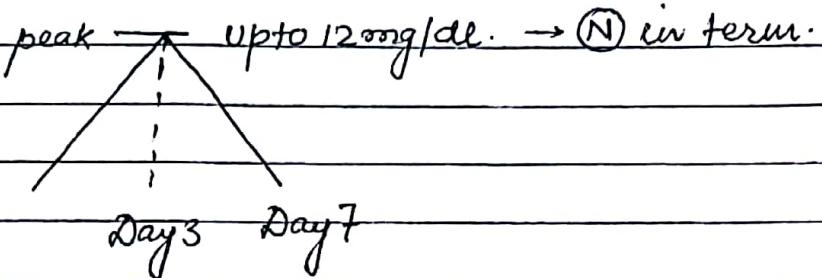
6 causes of physiological jaundice:

- ① Life span of RBC is less.
- ② Hematocrit more
- ③ Newborn deficient in γ -ligandin.
- ④ UDPglucuronyl-t deficient in newborn.
- ⑤ Excretion reduced.
- ⑥ Enterohepatic circulation ↑.

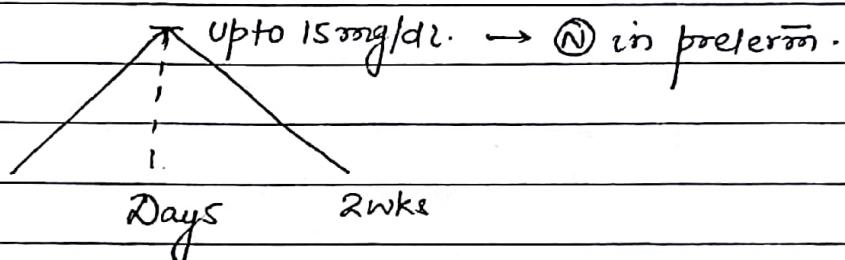


Physiological jaundice:

jaundice in term babies.



Preterm - have more jaundice.



Pathological jaundice:

- Hemolysis.

- M/c/c - Incompatibility Rh/ABO [O mother; baby A/B]
- RBC ~~membrane~~ membrane defect.
- RBC enzyme defect.

Def: jaundice in 24 hrs of life.

* Rh-ve mother; previous abortions
we take cord blood samples.

- Rh status of baby. \rightarrow Rh+ve
- Hb \rightarrow 10 mg/dl
- Bilirubin \rightarrow 5 mg/dl
- Peripheral Smear (P/S) +ve
- Direct Coombs test (DCT) \rightarrow +ve

\downarrow
means Severe hemolysis

Rx → Exchange transfusion at birth.

① jaundice in 24 hrs of life.

② Reaching 20mg/dl

③ Rate rise > 0.2mg/dl/hr.

④ Persisting beyond

- 1 wk term

- 2-3 wks preterm

⑤ Clay stools.

Phototherapy:

- Any jaundice on day 1 of life start phototherapy -
Serum bilirubin cut off.

Phototherapy

Healthy babies / Babies < R/F

Exchange transfusion

Healthy babies / Babies < R/F

Day 1	← Any visible jaundice →	260 (15)	220 (10)
Day 2	260 (15)	170 (10)	425 (25)
Day 3	310 (18)	250 (15)	425 (25)

Day 2	260 (15)	170 (10)	425 (25)	260 (15)
Day 3	310 (18)	250 (15)	425 (25)	340 (20)

Day 3	310 (18)	250 (15)	425 (25)	340 (20)
-------	----------	----------	----------	----------

R/F ① Gestation < 35 wks / wt. < 2kg.

② sepsis

③ Hemolysis

④ Asphyxia

⑤ Sick baby.

Principle:

- Structural isomerization

• Bilirubin → LUMIRUBIN

- Photo isomerization

• $4Z15Z \leftrightarrow 4Z15E$ (soluble).

- Minor pathway → photo-oxidation

• 40 cms away, falls @ 4-6 mg/dl/day.

- Phototherapy occurs at 425-475 nm of blue green light.
- Irradiance \rightarrow 6 micro watt/cm²/nm
- Intensive $>$ 30 micro watt/cm²/nm.

Q

AIIMS Nov. 2013

Which does not effect the efficacy of phototherapy?

- A) Types of phototherapy lamp.
- B) Skin pigmentation.
- C) Spectral radiance of incident light
- D) Initial bilirubin levels.

#

Complication:

- Hyperthermia; insensible losses
- Hypocalcemia
- Diarrhoea.
- Cover eyes & genitals
 - Retinal damage
 - Mutations
- Phototherapy is c/i in conjugated jaundice

↓
Bronze baby syndrome.
(Skin, Urine).

Exchange Transfusion:

- Double volume exchange transfusion.
- # Blood vol. of new born = 80 mL/kg
 - $\rightarrow 2 \times 80 \text{ mL/kg} = 160 \text{ mL/kg}$
- Transfuse fresh (< 7 days) whole blood.
 - Reduces bilirubin by 85%.

↓
Not 100% because bilirubin is also in tissue.

- Albumin ↑ ~~efficiency~~ efficiency.

Complications:

- Infection
- ACD (Acid citrate dextrose)

↓

Bicarbonates

1 mole of citrate → 3 mole of Bicarbonate.

- Metabolic alkalosis
- Hypokalemia, hypocalcemia.
- Old blood → Hyperkalemia, metabolic acidosis.

Persistent jaundice:

Cause — Hypothyrodesism

Breast milk jaundice

Hematoma (Cephalhematoma, IVH)

Clay colour stool (cholestasis)

Criggler - Najjar syndrome type II (milder form)
↳ deficiency of UDP-glucuronyl-*t*

Criggler - Najjar Syndrome type I — Very severe

↳ Absent UDP-glucuronyl-*t*.

Pathological jaundice

Breast milk jaundice

- Onset → day 14
- Some mother have pigment
- ↳ inhibit conjugation.
- Day 14 = 20-30 mg/dl
- May Kernicterus.
- Persists 4-6 wks.

Breast feeding jaundice

- Onset → Day 3.
- In primigravida.
- Starvation stimulates enterohepatic circulation.



Rx: Temporary interrupt
48-72 hrs.

Rx: Ensure feeding.

Meanwhile give
formula milk.

Q True about jaundice in newborn (neonates) is -

- A) Can be seen after Ventouse delivery.
- B) Physiological jaundice seen c in 48 hrs of birth.
- C) Increased conjugated bilirubin leads to kernicterus.
- D) Breast milk jaundice is maxⁱⁿ in 7 days of birth.

Neonatal cholestasis:

Neonate, jaundice & clay stools.

- Direct bilirubin $> 2 \text{ mg/dL}$ or $> 20\% \text{ total bilirubin}$

Medical - Common.

Neonatal hepatitis → CMV

Sepsis

Galactosemia

α -1 antitrypsin deficiency.

Neonatal hemochromatosis.

Surgical - Extra hepatic

biliary atresia (EHBA)

Rx - Kasai's surgery c in
8 wks of life otherwise

80% die.

↓

M/c/c of indication of
liver transplant in babies
→ EHBA.

Q Which is an ominous sign in a 10 day old newborn?

- A) Unconjugated hyperbilirubinemia
- B) Conjugated "
- C) Failure to gain wt.
- D) Doll's eye reflex
↳ Normal in 1st 10 days of life.

GGT → Gamma Gutaaryl transferase.



0312
Page

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Medical - Common

Surgical - EHBA

- GGT ten times higher in Surgical causes.
- Do USG → Shows Intra hepatic biliary radicle (IHBR) are dilated.
- Triangular cord sign

- HIDA nuclear Scan:

HIDA dye not seen in gut, even in delayed images; while in hepatites excretion of dye.

Best test - Liver biopsy.



Shows dilatation & proliferation of intra hepatic bile duct.

Before doing HIDA Scan we have to give phenobarbitone 2 to 3 days before test.

Gold Standard - Pre operative Cholangiography.

Alagille Syndrome -

- Cholestasis
- ~~AD~~ AD
- Bile duct paucity Syndrome.
- M/c heart disease → Peripheral pulm. stenosis.

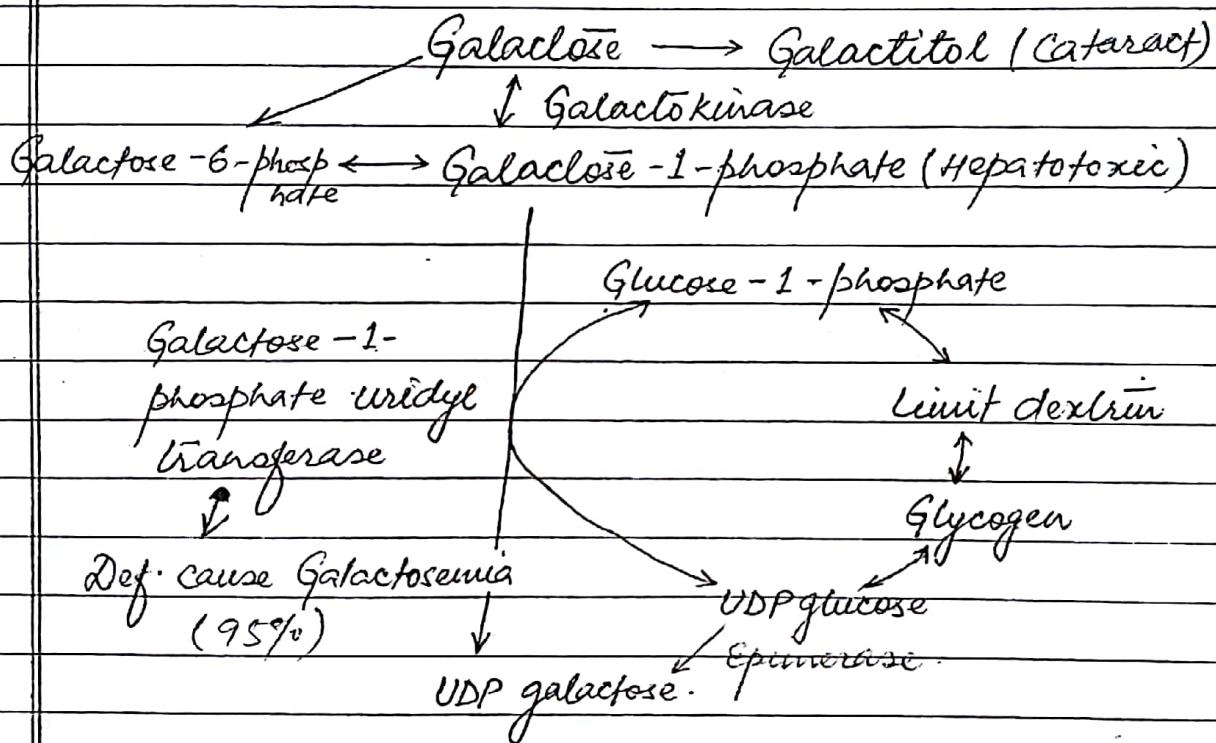
Rubella



Q. 1 month old child present with conjugated bilirubinemia & intrahepatic cholestasis. On liver biopsy staining with PAS, red coloured granules were seen inside the hepatocytes. Probable diagnosis is -

A) α -1-antitrypsin deficiency.
 B) Congenital hepatic fibrosis
 C) Hemosiderosis
 D) Wilson's disease.

CMV - Intracellular 'owl eye' inclusion bodies.



- He get jaundice, bleeding, hypoglycaemia, PTT.
- Galactitol cause cataract.
- Rx - lactose free milk.
- Galactokinase deficiency \rightarrow Only cataract
 No liver failure
- Fructokinase def. \rightarrow Causes Benign fructosuria
 \hookrightarrow No symptom.

Fructose-1-phosphate leads to liver failure.

Date 1/1
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Q. M/C/C of Neonatal Cholestasis:

- A) EHBA
- ~~B) Neonatal hepatitis~~
- C) Choledochal cyst
- D) Physiological

Q. Neonatal cholestasis seen in -

- A) Chronic hepatitis (>6 month)
- B) Hep. B
- ~~C) Galactosemia~~
- D) Rh incompatibility (cause ~~conjugated~~ unconjugated jaundice)

Q. Pregnant lady, HBsAg +ve, No jaundice.

↓

next step \rightarrow HBeAg

• HBeAg +ve (90%)

Carrier HBsAg \rightarrow Portal HTN

↓

Triad - Ascites

Splenomegaly
Varices.

• HBeAg -ve

anti HBe Ab \oplus

\hookrightarrow 10%

Immunization [HBIG - baby \in in 12 hrs of life

\downarrow HBV - baby in 24 hrs of life

prevent Vertical transmission



CDC guidelines:

Pricked by HBsAg \oplus patient



Are you immunized?



Anti-HBsAb titre

Good $> 10 \text{ mIU/ml}$

High risk $> 100 \text{ mIU/ml}$

Don't know titre \rightarrow Incomplete



HBs + HBsAg

If Good titre \rightarrow Nothing is to be done

Q.

New born, Respiratory distress:

Neonatal Seizures:



Neonatal hypoglycemia:

Causes:

- limited stores \rightarrow Preterm, IUGR's
- Stress \rightarrow Sepsis
- Polyhydramnios
- Galactosemia - liver failure.
- Low cortisol - CAH (Congenital Adrenal Hyperplasia)
- Hypopituitarism
- low ACTH; low LH/FSH \rightarrow Micropenis.
- Hyperinsulinism - In infants of diabetic mother.
 - \hookrightarrow Foetus of β -cell hyperplasia.

Foetus Blood glucose \downarrow
 \uparrow

Pederson's hypothesis

- Insulinoma; Nesidioblastosis
- Beckwith Wiedemann syndrome.
 - \hookrightarrow Hemihypertrophy of limb
 - \hookrightarrow Macroglossia
 - \hookrightarrow Risk of Wilms
 - \hookrightarrow Hyperinsulinism

Q. A term baby to a diabetic mother, few hours after birth - was lethargic & his blood glucose was 30 mg%. What should be done next -

A) Give 10% dextrose orally.

B) 10% dextrose i.v. - Bolus 2ml/kg → Glucose drip

C) Give expressed breast milk

D) DO exchange transfusion.

GIR
↓
6-8 mg/kg/min

Neonatal hypoglycemia:

Symptomatic

→ Bolus 2ml/kg 10% dextrose

if by Glucose drip

6-8 mg/kg/min

Asymptomatic

→ Blood glucose < 20 mg/kg

- ↓

Glucose drip (6-8 mg/kg/min)

→ Blood glucose = 20-45 mg/dl

↓

Breast feed; 1 hourly.

#

A baby on glucose drip & he gets seizures

how to ↑ glucose rate

- Up to 12.5% through peripheral vein.

Maxth glucose infusion rate (GIR) = 12 mg/kg/min.

Emergency drug = i.m. Glucagon.
for hypoglycemia

↓
Glycogenolysis
+ Gluconeogenesis.



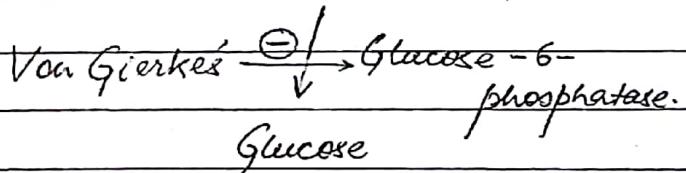
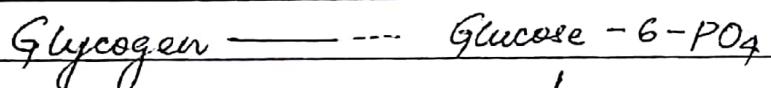
Q. Glucagon is effective for Mx in persistent hypoglycemia in all except.

- A) Large date for baby.
- B) Neuridioblastosis
- C) Galactosemia
- D) Infant of diabetic mother.

Q. 1 yr old, hypoglycemia & hepatomegaly
No jaundice.

Hypoglycemia doesn't respond to glucagon.

Δ → Von Gierke's (Glycogen storage ds - I)



Types of Glycogen storage disorder:

V = Von Gierke's → Liver primary

P = Pompe's ds → Heart primary; Cardiomegaly.

C = Cori → Debranching enzyme. Large QRS complexes.

A = Anderson → Branching " deficiency

M = Mc Ardles → Muscle phosphorylase deficiency.

Harding =

Ton =

jaundice never occurs in Glycogen storage ds.

Rx pompe's: Enzyme replacement therapy.

Enzyme absent in pompe's → lysosomal α -1,4-glucosidase
- Also called Acid & neutral maltase.

Muscle affected in GSD:

Calcium:

(N) S. Ca^{2+} \rightarrow 9-11 mg/dl.

Neonatal hypocalcemia:

- S. Ca^{2+} $< 7 \text{ mg/dl}$
- ~~base~~ Best index of body calcium

Ionized $< 4 \text{ mg/dl}$ (or) $< 1 \text{ mmol/L}$

- Tetany is rare in infants.

- Tremors, seizures, jitteriness.

↳ Tremulousness \leq is stimulus sensitive.

↳ Can stop on passive restrain.

Early Hypo Ca^{2+}

Causes - Prematurity
Asphyxia

↳ Infant of DM mother.
↳ Test:

Blood glucose

+ S. Ca^{2+} & S. Mg^{2+}

Late Hypo Ca^{2+}

Cause - Feeding \bar{c} phosphate rich milk (cow milk)

Good Ca^{2+} Supplement ($\text{Ca}^{2+}/\text{P} > 2$)

100 ml

Ca

PD₄

Cow milk

118 mg

100 mg

Breast milk

34 mg

15 mg

Advantages of Breast milk:

- protects against late onset hypocalcemia.
- protect against pneumonia

NEC (Necrotizing Enterocolitis).

Allergy, Eczema, asthma.

Rota virus diarrhoea.

Bronchiolitis (IgA-RSV)

Q. Milk deficient in:

a) Iron & Vit. C

Q. Breast milk has enough iron & Vit. C for 6 months.

∴ Scurvy never occurs in ^{1st} 6 months of life.

Q. If baby on exclusive breastfeed for 2 yrs.

Iron deficiency anaemia.

Q. APP recommends Vit. D to all infants - Vit. D drops.

- RDA Vit.D infant 400 IU/day
- Breast milk 25 IU/L.

M/c/c of HypoCa²⁺ in infants = Maternal deficiency of Vit.D.

25[OH] Vit.D = Status.

Infant of DM mother:

- Can be stillborn; preterm
- Macrosomia
 - ↳ Bcoz of Hyperinsulinism
- Linear growth in utero depend upon insulin & or Insulin like growth factor.
- LUGR → White's classification class F/R
- mother → Placental vasculopathy.
- Hypoglycemia
- Hypo Ca^{2+} , Hypo Mg^{2+}
- Neonatal jaundice
- Polycythemia → Renal Vein Thrombosis (RVT)

Q. Atq. Not seen in infant of diabetic mother:

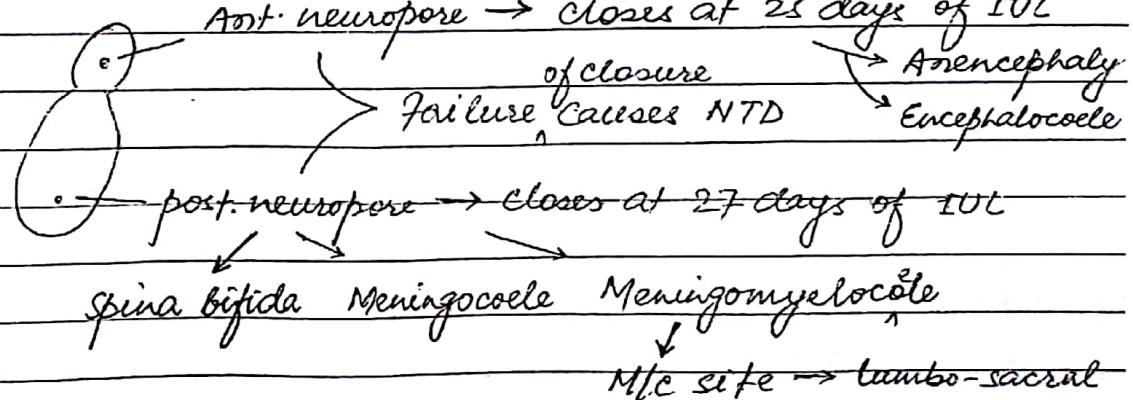
↓
Hyperglycemia.

Anomaly:

(M/c)- CVS - 8.5% [VSD; HOCM] Asymmetrical septal hypertrophy

- Neural tube defect → 5% hypertrophy
- Lazy left colon syndrome → Pseudoobstruction of colon
- Sacral agenesis/Caudal regression syndrome
 - ↳ Most specific.

NEURAL TUBE DEFECT (NTD)



Anecephaly:

- Ant. neuropore fails to close.
- No brain, absent of part of hind brain.
- Earliest abnormality diagnosed by USG (10-12 wks) of gestation.
- Most severe NTD
- Don't resuscitate
- Mostly post-term.

Herniation of brain tissue - Encephalocele.

Lumbo-sacral myelomeningocele:

Complication:

- Paraplegia/Paraparesis
- Neurogenic bladder → CKD.
- Constipation (Severe)
- Associated hydrocephalus. (obstructed)

Bcoz of Arnold-Chiari type II malformation

¶ MRI Brain

Ruptured myelomeningocele:

Best test - Blood culture.

- Cover w/ Normal saline soaked gauze.
- 95% of neonatal meningitis have leukemia.
- M/c/c of meningomyelocele - Folic Acid deficiency.

Folic acid deficiency:

- -1/+3 months conception - start folic acid.
- 400 mcg given.

Recurrence:

1 child - 3.5% chance

2 child - 10%

3 child - 25%

To prevent recurrence → Folic acid 4mg

↓
reduces risk by 75%.

△: ① USG

② Amniocentesis

↳ Acetylcholinesterase

& α -fetoprotein are markers.

α -fetoprotein in mother serum is marker of Neural tube defect.

Resuscitation:

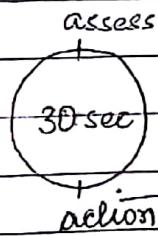
NRP Guidelines 2015:

T = Temp

A = Airway: position neck, suction

B = Breathing

C



Indication of Bag & mask \bar{c} 100% O₂:

- Apnea/gasping ~~after~~ after initial steps.
- HR < 100/min after 30 sec PPV.
- Central cyanosis despite 100% O₂.
- Chest compression if HR < 60/min, falling after 30 sec PPV.
- Chest compression: Bag mask = 3:1

In 1 minute = 120 events

90 Chest compression & 30 Bag & mask.

Compression to Ventilation ratio:

- Children/Infant - Single rescuer 30:2
2 rescuer 15:2
- Adults - 1 or 2 rescuer 30:2

CPR sequence \rightarrow CAB

Drugs for resuscitation:

- 1) 0.9% NaCl 20ml/kg bolus \rightarrow shock
- 2) 1:10,000 epinephrine 0.1-0.2ml/kg

↓

if HR = 0 or falling.

- 3) I.V. NaHCO₃ - documented metabolic acidosis.
- 4) I.V. Naloxone - mother opioid addvt.

Targeted preductal SpO₂ after birth:

1 min = 60% - 65%

2 " = 65 - 70 %

3 " = 70 - 75 %

4 min = 75 - 80%

5 min = 80 - 85%

10 min = 85 - 90%

GENETICS

   → Heterozygotes for AR carrier

 → Carrier for sex-linked recessive

 → Death

 → Abortion or stillbirth
Sex unspecified.

 → Proband.

  → Consanguineous
marriage

  → Dizygotic twin

  → Monozygotic twin

  → azoospermia

 → Endometriosis

  → Infertility

   → Adopted in

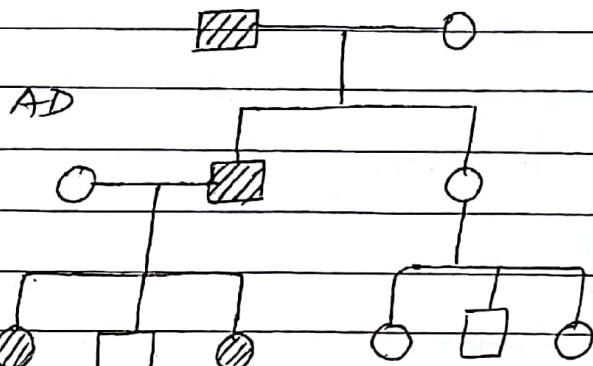
   → Adopted in

  → No children for choice
or reason unknown

 → Vasectomy

 → Tubal

  → Divorce.



Autosomal dominant:

D = Dystrophy myotonie → distal myopathy.

O = Osteogenesis imperfecta

M = Marfan Syndrome

I = Intermittent porphyria

N = Noonan Syndrome → Turner phenotype XX & XY.

A = Adult PKD, Achondroplasia

N = NF (Neurofibromatosis)

T = Tuberculous sclerosis

VH3 = Von Willibrand Syndrome

Huntington's chorea

Familial Hypercholesterolemia

Heredity spherocytosis.



Turner Syndrome

- 60% case - XO
- Webbed neck
- Cystic hygroma
- Lymphedema of hand & feet
- Primary amenorrhea during puberty.
- Streak ovaries
- Cubitus valgus
- MR rare.
- M/c heart ds - Bicuspid aortic valve & out stenosis
↳ half to one-third.
- 20% coarctation.
- Girls infertile

Noonan Syndrome

- AD
- XX = XY
- 25% MR
- Valvular pulm. Stenosis. HOCM
- ASD.

- Girls are mostly fertile.
- Boys - Cryptorchidism.
- Clotting factor deficiency.

Q. Chance of child being not affected if both parents are affected c^r Achondroplasia is -

A) 0%

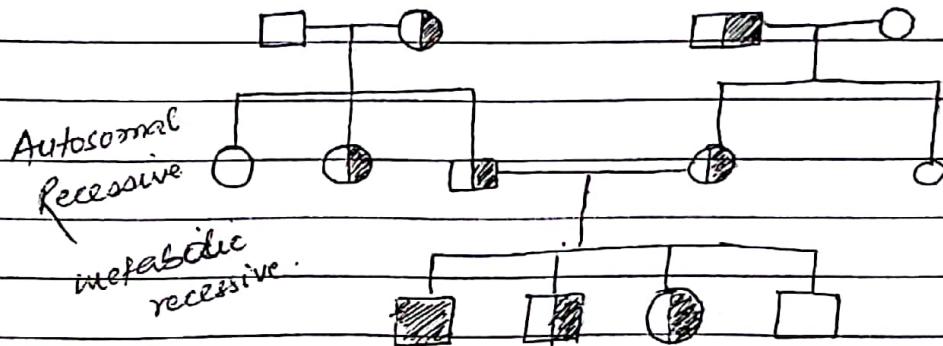
B) 25%

C) 50%

D) 100%

$AAC \times AAC$

(AA) AAC ACA ACA^c





Autosomal Recessive:

Cystic Fibrosis

• δ , AT deficiency

Wilson's ds

Haemochromatosis

Friedreich's ataxia

Gaucher's ds

Niemann's pick ds.

Tay Sach ds.

Hurler's Syndrome.

Mucopolysaccharidosis (MPS)

- Child c noisy breathing
- Coarse facies.
- Ab (n) accumulation of glucoseaminoglycans
- Gargolism
 - ↳ Chronic rhinitis.

Mucopolysaccharidosis type I \rightarrow k/a HURLER'S

Syndrome



deficiency of L-iduronidase.

Rx - Enzyme replacement therapy.

HURLER

- MPS - I
- AR
- Corneal clouding

HUNTER Syndrome.

- MPS - II
- XL recessive
- Cornea clear.

Enzyme Replacement therapy:

1st to be treat → ① Gaucher's ds

(β glucocerebrosidase)



Company name — GENZYME
CER-zyne.

② Pompe's

③ Hurler (MPS-I)

④ MPS-VI (Maroteux-Lamy)

⑤ X-linked recessive Fabry's

(CNS & kidney problem)



Lysosomal disorder.

Cherry red spot macula seen.

Gene therapy:

1st to be treated by Gene therapy → X-linked recessive

Severe combined immunodef. (SCID)

(Adenosine deaminase deficiency)

British guy British lady

Chance of child having cystic fibrosis

UK/ Europe → $\frac{1}{25}$ carrier cystic fibrosis.
Ashkenazi Jews

$$\frac{1}{25} \times \frac{1}{25} \times \frac{1}{4} = \frac{1}{2500}$$

$AAC \times AAC$

$(AAC) \cdot AAC AAC AA$
 $\frac{1}{4}$

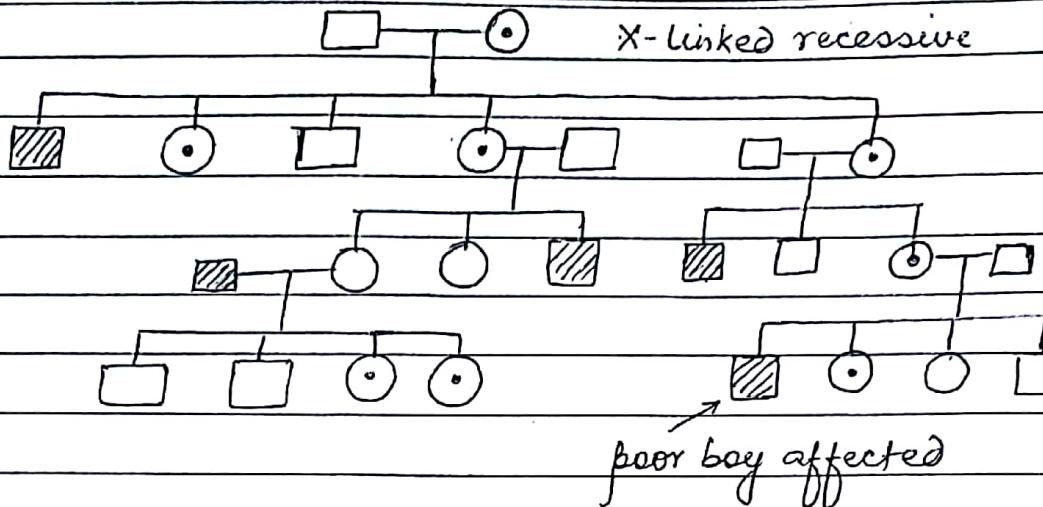
British guy British lady (brother died of cystic fibrosis)

Chance of child to have cystic fibrosis

$$\frac{1}{25} \times \frac{2}{3} \times \frac{1}{4} = \frac{1}{150}$$

$AAC \times AAC$

~~$(AAC) \cdot AAC AAC AA$~~



XX^C XY
 XX XX^C $X^C Y$ XY

X-linked Recessive: (Poor ~~boys~~ boys)

- Duchenne muscular atrophy. (M/c hereditary Neuromuscular disorder)
- Hemophilia A & B.
- G-6PD deficiency.
- Wiskott-Aldrich syndrome.
- Colour blindness.
- Lesch-Nyhan syndrome.
- Chronic granulomatous disease.

Duchenne muscular hypertrophy (DMH):

- Pseudo hypertrophy of calf muscle bcoz of fat deposition.
- Proximal muscle weakness
- Gower's Sign \rightarrow Not specific.

△ \rightarrow CPK = 10,000 IU

Valley Sign \Rightarrow Hypertrophy of Supraspinatus

↓ Atrophy of Infraspinatus

Also seen in DMH.

↳ Specific for boys who don't have calf hypertrophy.

Human Genome:

- 30,000 genes.

Largest gene - Dystrophin

↳ Skeletal muscle

↳ Heart - Cardiomyopathy.

↳ Brain \rightarrow 1/3 cases MR.

Boys - Duchenne - die teens due to recurrent chest infection.

Duchenne \rightarrow XLR, 1/3 de novo mutation.

Becker's dystrophy:

- Similar to Duchenne
- Mild form & present late.
- X-linked recessive.

Wiskott-Aldrich:

- X-linked recessive
- Eczema
- Thrombocytopenia
- Immunodeficiency.

Chronic granulomatous disease -

- X-linked recessive
- Immunodeficiency.
- ~~Thick~~ NADPH oxidase deficiency
- Dx \rightarrow NBT dye test

Lesch - Nyhan Syndrome:

- X-linked ~~recessive~~ recessive
- Purine defect
- HGPRTase deficiency.
- Hyperuricemia $> 6.5 \text{ mg/dl}$
- Self-mutilation \rightarrow nose, palate, fingers.



Q. A mentally challenged child with dysphagia & opisthotonic spasms. He is also having choreoathetoid movements & self mutilation behaviour. There is a family history. Which of the following investigation is suggested?

- A) Serum uric acid. → Lesch Nyhan Syndrome.
- B) S. ALP
- C) S. LDH
- D) Lead level in blood

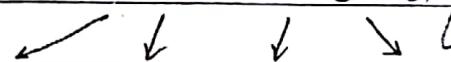
Q. A male child with Fanconi Syndrome with nephrocalcinosis have a variant of dent disease. All true except

- A) Hypercalcioria → 24hr urine $\text{Ca}^{2+} > 4 \text{ mg/kg}$
- B) Proteinuria → LMW (β_2 microglobulinuria)
- C) Similar presentation in father
- D) Rickets

Urolithiasis

Nephrocalcinosis

Fanconi Syndrome - Proximal tubular defect



Na^+ HCO_3^- PO_4^{2-} AA, Glucose

60% >85% >88% 100%

[Lowe Syndrome] - X-linked recessive.

↳ Oculo-cerebro renal gene. (OCRL gene).



Congenital cataract Microcephaly Fanconi's
" glaucoma MR Syndrome.

Fructose-1-phosphate] proximal tubular toxic
Glucose-1-phosphate

Franconis Syndrome:

- Genetic

↳ X-linked recessive - lowe

XLR Dent

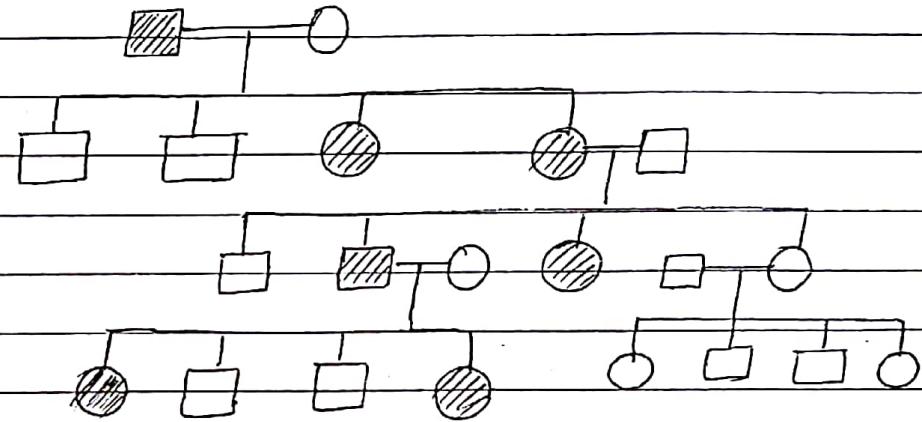
AR

↳ Cyblosis

- Metabolic - Galactosemia, HF-1

Tyrosinemia (R-Nitroscine).

- Acquired - expired tetracycline



X-linked dominant father to all daughters - none son

X-linked dominant: Males are more severely affected

- Familial hypophosphatemic rickets.

- Urea cycle defect due to OTC deficiency.

- Incontinentia pigmenti [Only seen in girls.]

- Rett's syndrome.

bcz boys die.

(lethal in male fetus).

Rett's Syndrome:

- Pervasive ~~perma~~ developmental disorders.
 - ↳ Autism (Common in boy ~~girl~~ < Syn)
 - Asperger
 - Rett's

Q. Not seen in autistic ~~disorder~~ disorder -

- A) Social avoidance
- B) Visual impairment.
- C) Interest in one self
- D) Introvert person.

Asperger's Syndrome: Common in boys

Very good IQ.

Rett's Syndrome - Common in girls

X-linked dominant.

Normal till 6-18 months

Decrease in head growth.

Microcephaly; MR

Hand movements

Repetitive behaviour

MeCP2 gene mutation

↳ Macrocephaly not seen.

↳ Abnormal dendritic morphology in
cortical pyramidal cells
(postmortem brain biopsy).

↳ Seizures.

Cause of death → Arrhythmias (cardiac)

↓
Sudden death.

Ornithine Transcarbamoylase deficiency:

Orotic aciduria → URACIL

→ X-linked disorder.

- The mother of these child have also high Uracil level in urine.

Deletions:

Major deletion

→ Cri-du-chat

↳ Deletion of 5p chr.

- Cry like cat due to absence of larynx.

Microdeletion

→ Williams (7q23-) Syndrome

→ Prader-Willi Syndrome

(11q11-13-)

→ Di-George's Syndrome (22q11-)

Δ → by FISH.

Williams Syndrome:

Supravalvular aortic stenosis

→ peripheral pulmonary stenosis.

- Hypercalcemia

- Elf in facies.

Di-George's Syndrome:

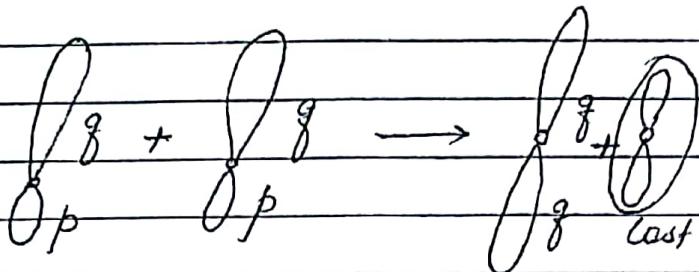
- Hypoplasia of 3rd & 4th pharyngeal pouch.

- Absent Thymus

Parathyroid.



TRANSLOCATION:



- Unbalanced

- Robertsonian Translocation:

Translocation b/w two Acrocentric chromosome.

M/c/c (Genetic) of MR = Trisomy 21 (Down's Syndrome)

Extra chromosome is of females.

Trisomy 21 - In 95% cases: Maternal meiotic non-dysjunction.

- 3-4% - Robertsonian translocation

- 1-2% Mosaic 47/46

carrier of RT look normal

but they can have abn children.

MOSAICISM:

A single zygote giving rise to different cells.

CHIMERA - different zygote giving rise to different cells
 ↳ Rare in humans.

- Mosaicism Seen in humans

- 1-2% are down's

Klinefelter Syndrome → 80% XXY; XY/XXY;
 XY/XXXYY.

Turner's Syndrome:

Cytogenetics

60% 45X0

15% Mosaic XX/X0

10% Isochromosome Xq or Xp. — Mentally Retarded.

10% 46X deletion.

5% Mosaic X0/XY → Risk of Gonadoblastoma.

Loss of one arm & duplication of other



Isochromosome

Mosaicism ↗ Somatic — Not transmitted

Germline — Transmitted

↳ Blood DNA is normal.

eg: ~~Osteogenesis~~ Osteogenetic imperfecta.

Q. Couple has two children with tuberous sclerosis. On detailed clinical & lab evaluation (including molecular studies) both parents are normal. ↗ one of the following explains the 2 affected children in this family —

A)

B)

C)

✓ D) Germline mosaicism.

Maternal inheritance → Mitochondrial.

Mitochondrial inheritance:

- MERRF (Myoclonic epilepsy & red ragged fibres)
- Mitochondrial encephalopathy, stroke-like episodes, & lactic acidosis (MELAS)
- Leber hereditary optic neuropathy (LHON)



- Leigh disease
- Kearne - Sayre Syndrome (KSS) (ophthalmoplegia)
- NARP (Neuropathy, Ataxia, Retinitis pigmentosa)
- Chronic progressive ophthalmoplegia
- Pearson's Syndrome : Panhypoproteinemia
+ Pancreatic insufficiency

Anticipation : Severity of genetic disorder ↑ in every successive generation.

More repeats — more problem.

eg: All Tri nucleotide repeat disorders

- Fragile X — CGG repeats
- F. Ataxia — GAA "
- Myotonic dystrophy — CTG, CCTG
- Spinobulbar muscular dystrophy — CAG
- Huntington's — CAG
- Spinocerebellar ataxia — CAG/CTG.

Fragile X : genetic

- End inf. cause of MR in boys.
- X-linked
- Large face
- Large ears
- Prominent jaw
- Large testis..
- CGG repeats > 1500 repeats

Genomic imprinting:

Chr. 15q • 11-13 deletion.

↳ Paternal inheritance cause Prader-Willi Syndrome

↳ Maternal " "

Angelman "

- Differential expression of genetic appearance depending upon parents.

Prader-Willi Syndrome

- Severe neonatal hypotonia.
- Obesity
- ~~Small~~ Small hands & feet. (Rx - GH treatment)
- Unusual behaviour
- MR.
- High Ghrelin → So obese. (Rx - Anti Ghrelin)

Angelman Syndrome:

- (R) at birth
- K/A happy puppets.
- Subsequently develop seizures
- MR
- Ataxia

↓
appetite hormone
for fundus.

Genomic imprinting → Prader Willi Syndrome (70%)

↓

Almond shaped eyes.

② Angelman Syndrome (70%)

③ Neonatal DM

④ Beckwith Wiedemann

⑤ Seckle

⑥ Temple

⑦ Wang

⑧ Pseudohypoparathyroid Ib.



Q. Father carrier of cystic fibrosis - AA^c
 Mother - $(N) \rightarrow AA$



Yes there is a chance of cystic fibrosis in child
 AA^c



Uniparental disomy.

- In some cystic fibrosis
- In some sickle cells.
- 30% Prader Willi - Unimaternal disomy.
- 5% Angelman - Unipaternal disomy.

PRADER WILLI :

70% - Paternal inheritance deletion.

30% - Unimaternal disomy

↳ Maternal silencing gene.

Congenital Heart de:

NADA'S CRITERIA:

1 Major & two minor criteria at least.

Major:

- ① Systolic murmur grade III or more ass. c thrill.
- ② Diastolic murmur
- ③ Cyanosis (Central)
- ④ CHF

Minor:

- ① Systolic murmur < grade III
- ② Abn S₂
- ③ Abn ECG
- ④ Abn X-Ray
- ⑤ Abn BP

A₂ ----- P₂ Expire

A₂ ----- P₂ Inspire

ASD - Wide & fixed S₂.

A₂ ----- P₂ Expire

A₂ ----- P₂ Inspire.

- Volume overload in right ventricle.

VSD - Wide & Variable S₂.

A₂ ----- P₂

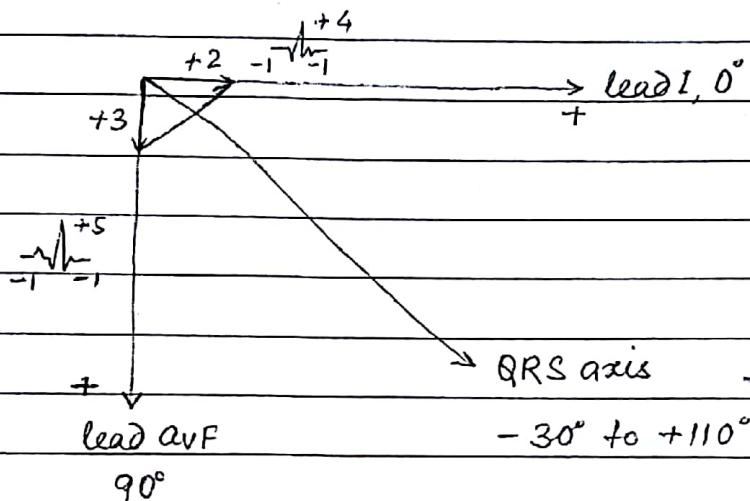
A₂ ----- P₂



TOF - Single S_2 (A_2).

All newborn has RVH & RAD.

- Axis like adult $> 1\text{st month life}$
- T-wave V_1 ; V_3R ; V_4R
 - 'up' first 48 hrs.



- Negative after 48 hrs
- Never be +ve $< 6\text{ yrs}$
- $> 6\text{ yrs} \rightarrow$ positive.

Prevalance of CHD:

- Prevalence = 0.8 - 1%

Recurrance = 2-6%

M/C CHD = VSD (30-35%).

2nd M/C CHD = ASD (Secundum) - 6-8%

> PDA (6-8%)

> Co-arctation of Aorta (5-7%)

> TOF (5-7%)

> Pulm. Valve stenosis (5-7%)

> Aortic * * (4-7%)

M/C Syndrome in CHD = Down's Syndrome.

Down Syndrome:

M/C → Complete AV septal defect (CAVSD) /
AV canal defect / Endocardial cushion defect
Ostium primum ASD.

(37%) — M/C/C of death.

- VSD (31%)
- ASD (15%) → Secundum
- Partial AV septal defect (PAVSD) — 6%
- TOF → 5%
- PDA → 4%
- Miscellaneous → 2%

Turner's → half to one third bicuspid aortic valve;
20% coarctation.

Noonan's → Turner's phenotype.

AD; XX = XY

Valvular pulm. stenosis.

HOCM; ASD.

Rubella → PDA; Peripheral pulm. stenosis; VSD.

Rubella

Alagille Syndrome
William's "

Maternal lithium → Ebstein's anomaly.

Maternal mumps → Endocardial fibroelastosis/
LV obstruction newborn.

Maternal penicillamine → Cutis Laxa.



Maternal SLE << Maternal Sjogren Syndrome

- New born complete heart block.

pacemaker insertion (anti ~~Ro~~ Ro).

Maternal warfarin:

Chondrodysplasia punctata.

Maternal thalidomide: Phocomelia.

Foetal circulation:

1 Umbilical Vein (left)
2 umbilical artery.

Ductus venosus →

P_{O_2} in umbilical vein = 30-35 mm of Hg

P_{O_2} of IVC in fetus = 28-30 mm of Hg.

As soon as child takes his 1st breath

- Umbilical artery constricts.



U. vein closes



Ductus venosus closes.

Ductus arteriosus:

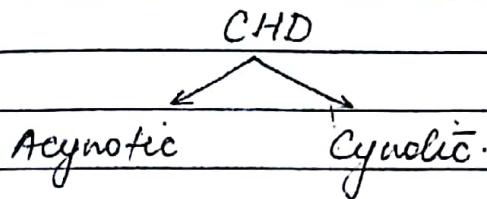
- Physiological closes in 10-15 hrs.

- Anatomical → 10-21 days.

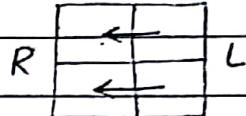
Foramen ovale:

Functionally closes by 3 months.

Anatomically 10-15% open.



Acyanotic : $L \rightarrow R$ Shunt
 ASD, VSD, PDA.



Pulm. blood flow \uparrow (Plethora)

↳ lung vascularity is good.

C/F: Failure to thrive

Recurrent pneumonia

Feeding diaphoresis

CHF in 6-10 wks of life.

Suck-rest-suck cycle

\uparrow sweating (d/t sympathetic stimulation).

Tachycardia] in every CHD.

Cardio megaly]]

Cyanotic

PBF

↑

↓

Plethora

Oligemia

- Persistant Truncus

arteriosus

(Cyanotic + Recurrent pneumonia)

- dTGA + VSD

- TAPVC

Massive

Cardiomegaly.

· Ebstein's

(Box/Ballot heart)

physiology.

① Heart

Single S₂

F · TOF

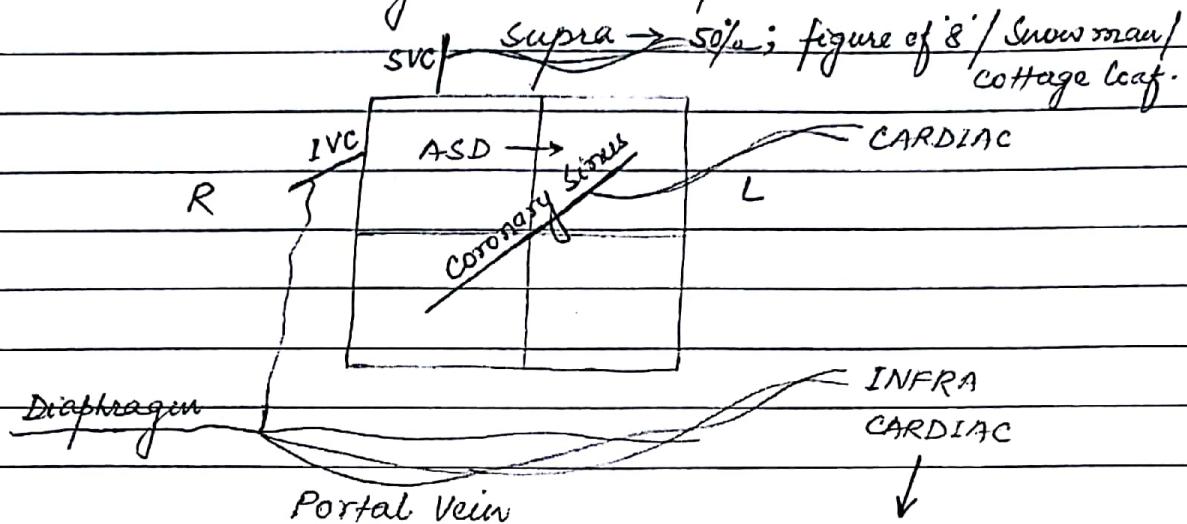
L · DORV + PS

O · dTGA + VSD + PS

T · Single Ventricle + PS.



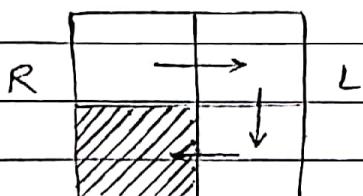
TAPVC (Totally anomalous pulm. venous connection):



More severe TAPVC -

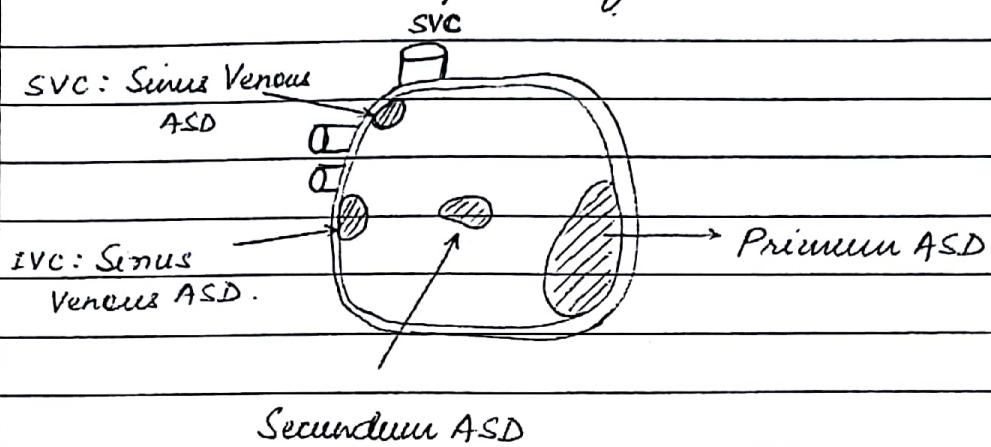
- Cyanosis at birth.
- Obstructive pulm. venous hypertension.
- Ground glass : Kerley B lines.
- Q - Worsen by PGE1 infusion.
- Only pediatric cardiac Sx emergency.

■ Tricuspid Atresia:



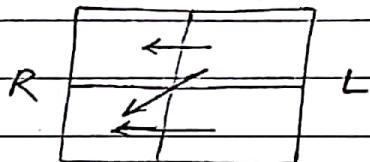
- Cyanosis + ↓ PBF
- + LVH
- LAD

ASD (Atrial Septal defect):



M/C syndrome ass. = Down's Syndrome.

Endocardial cushion defect.



- Very large L-R shunt.
- PBF↑
- Pulm. plethora

ASD Syndrome:

- HOLT ORAM : • Familial, AD,
 - ASD Secundum; VSD; 1 degree block;
 - AF + Bone defect (Absent Radius).
 - Distally placed thumb / Rudimentary thumb / Triphalangeal thumb.
 - A/K/A Hand-heart Syndrome.
 - TBX5, mutation → Pleiotropy.
 - ↳ Common transcriptional factor for hand & heart.



Absent Radius associations:

- ECG (Holt-Oram)
- Platelet (Thrombocytopenia ; AR).
- Bone marrow biopsy (Fanconi's anemia)



Congenital aplastic anemia

- Rarely Karyotyping (Edward Syndrome)

ASD Syndromes:

- Down's Syndrome
- Holt Oram
- Lutembacher \rightarrow ASD + Mitral Stenosis
- Ellis Van Creveld \rightarrow ASD + polydactyly.

ASD Secundum:

- Child \rightarrow asymptomatic, wide & fixed S₂.
- ECG \rightarrow RAD (Right axis deviation).
- In adult life \rightarrow Complications
 - RV failure
 - Arrhythmias; AF \rightarrow CVA
 - Reversal; R \rightarrow L (Eisenmenger Syndrome)

Natural history:

If < 3 mm \rightarrow close itself

> 8 mm \rightarrow unlikely to close; Require Sx.

Indication of Sx in ASD Secundum:

- All symptomatic.
- Q_p / Q_s > 2 ; even if no symptoms.
 \hookrightarrow Pulm blood flow / Systemic blood flow.

Q. Least chances of Infective endocarditis is seen in ?

- A) Small VSD \rightarrow M/c/o of IE
- ~~B) Small ASD secundum \rightarrow Rare~~
- C) Mild AS
- D) Mild AR.

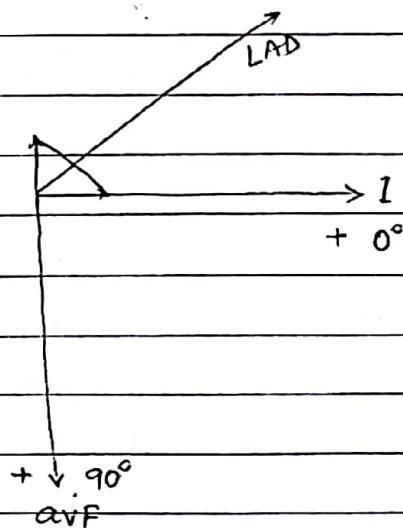
ASD Secundum doesn't require any prophylaxis t/t before going to Sx.

ASD primum + Mitral Regurgitation:

- Wide & fixed S₂ +
 $S_1 = S_2$ apex \rightarrow axilla & back.
- 6 - 10 wks presents in CHF.
- Conduction defects; ECG \rightarrow LAD.
- Common in Down's Syndrome baby.

LAD d/t endocardial cushion defect.

Down's
AVSD





VSD (Ventricular Septal defect):
70% → Perimembranous

Small VSD

- Root of aorta $< \frac{1}{3}$
- $< 3 \text{ mm}$

Medium VSD



Large VSD

- $> \frac{1}{2}$

- Called Maladie de Roger's defect.
- Loud murmur (S₁)
(Pansystolic)

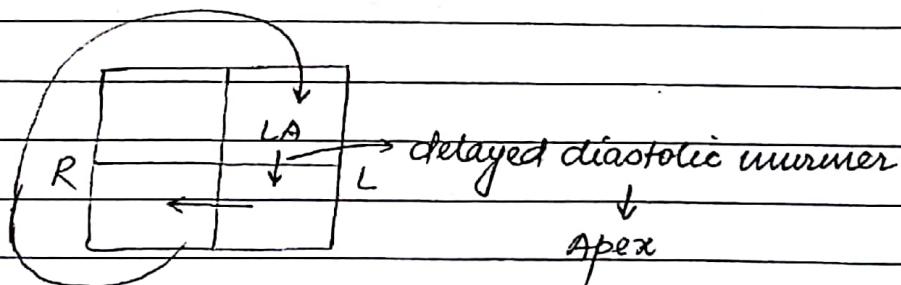
lower left Sternal
border.

- Asymptomatic

CHF at 6-10 wks of life

- No murmur/
- Ejection systolic murmur.
(b/w S₁ & S₂).

In medium to large VSD, Left Atrium enlarges first.
In Small to medium VSD, left Ventricular hypertrophy.
due to blood overload.



Natural course of VSD closure:

- 80% perimembranous
- 50% muscular VSD's close.
- By 4 yrs.

Indication of Sx in VSD:

- Failure of Medical therapy.
 - ↳ Digoxin
 - ↳ Diuretics
 - ↳ Dilators
 - ↳ ACE i (Remodelling of heart).
even $Q_p/Q_s > 2$; if no symptoms.
- Swiss cheese VSD. (Multiple; apex)
- Supracrestal (outflow)

Large L \rightarrow R shunt



Pulm. blood flow ↑



Irreversible changes pulm. microvascular.

[Pulm. HTN]



RV pressure ↑; RVH



Reversal; R \rightarrow L (Eisenmenger Syndrome).



Sx is CI !

Differential clubbing: Toes > fingers



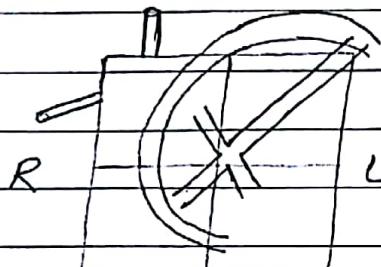
PDA + Reversal.

(also differential cyanosis).

Down's Syndrome baby c Endocardial cushion defect undergoes Eisenmenger Syndrome.



Ductal dependent lesion:



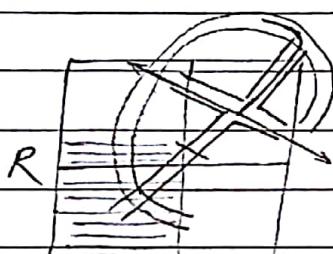
S/E - Apnoea.
- HLHS \Rightarrow Rx - (PGE₁) infusion.

- Critical AS
- Preductal coarctation
- Interrupted aorta
↳ Shock.

I > Systemic blood flow dependence.

II > Pulm. blood flow dependence.

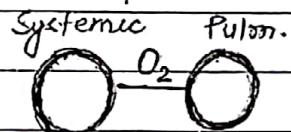
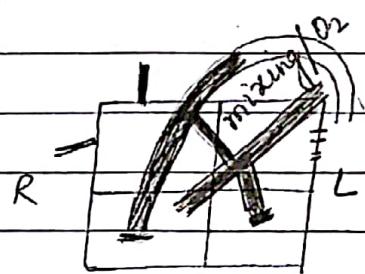
- Tricuspid Atresia
- Pulm. Atresia



↳ Central cyanosis.

In emergency cond'n \rightarrow PGE₁ infusion.

III > d-TGA



Rx - PGE₁ infusion

d-TGA \rightarrow dependent for mixing.

Truncus Arteriosus is Ductal independent.

Emergency Sx in TGA \rightarrow Rashkind's
Balloon atrial septostomy.

Definitive Sx \rightarrow Jatene's Arterial Switch.
↳ Best time: c in first
2 weeks of life.

M/c Cyanotic Heart de:
 Overall - TGA

In Infants - d-TGA (< 1 yrs)
 > 1 yr - TOF

Hyperoxia test in cyanotic newborns -

- 10L O₂ to cyanotic newborn
- $pO_2 > 150$ mm Hg \rightarrow excludes heart de.

① 50% d-TGA + VSD \rightarrow Mild

② d-TGA + intact septa (complete d-TGA) - Birth.

③ d-TGA + patent foramen ovale \Rightarrow At birth.

① < ③ < ②

TOF (Tetralogy of Fallot):

M/c cyanotic heart de beyond infancy.

- ① Narrowing of the pulm. valve (infundibular).
- ② RV hypertrophy.
- ③ Overriding of aorta over VSD.
- ④ VSD - opening b/w left & right ventricles.
 \quad (perimembranous).

Pink child become a blue TOF.

- Cyanosis; clubbing.
- Polycythemia \rightarrow R/F of Renal vein thrombosis (RVT).
- Hematuria, ~~microthrombi~~. Flank mass, Anemia.
- Infant - Cyanotic/tet/hyperpneic spells \rightarrow older squat
- Complications:
 \quad ① < 2 yrs = thrombosis



⑪ > 2 yrs = Brain Abscess.



in the territory of middle
meningeal artery
(Parieto - Temporal)

Infants → Cyanotic/ tet/ hypoxic spells - older squat

(R: knee chest position; - Systemic Vascular Resistance falls.

i.v. Ketamine; - R → Shunt ↑ - Murmur disappears.

Phenylephrine) - PO₂ falls (R: O₂; i.v. NaHCO₃).

- Respiratory centre + + + (R: S/c Morphine)
- Dynamic Pulm. stenosis (R: i.v. Propranolol)
- Crying (Bcoz of ↑ venous return).

In TOF - loud Holosystolic murmur at the left
2nd - 3rd ICS due to pulm. stenosis.

Palliative Shunt

- Modified Blalock - Taussig → Subclavian
to Pulm. artery
(opposite to Aortic Arch.)
- Waterston
- Pott's (Descending Aorta to left pulm. artery)



25% TOF ⇒ has Right aortic arch.



M/c c 50% of Truncus arteriosus

Pentalogy = + ASD

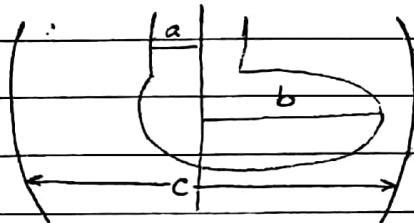
Trilogy = Overriding of aorta absent; VSD absent; ASD present.

CHF never seen in TOF

Cardiomegaly never seen in TOF.

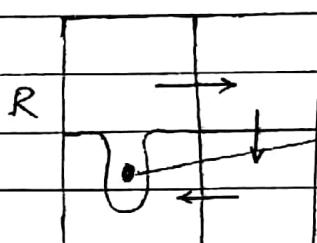
Cardiothoracic ratio > 0.6 infant

> 0.55 in older infant.



"Boot shaped heart".
"Coer en Sabat".

Ebstein's anomaly:



• Cyanosis \Rightarrow ↓ PBF

• Maternal lithium

→ Atrialisation of RV.

Pressure = RA

Ventricular = ECG

disconcordance b/w pressure & ECG.

• Intracardial ECG helpful in Δ.

• Systolic; diastolic murmurs.

Quadruple rhythm

• Box of Balloon; Massive heart.

• SVT; WPW Syndrome.



Q A neonate presents with recurrent abdominal pain, restlessness, irritability & diaphoresis on feeding. Cardiac auscultation reveals a non-specific murmur. He is believed to be at risk of MI.

The most likely Δ is —

- A) VSD
- B) ASD
- C) TOF

\checkmark D) Anomalous origin of the left coronary artery
= ACCAPA

- Q wave in Lead I; aVL
- LAD branch absent.
- Ischemic LV \rightarrow Anterolateral MI.

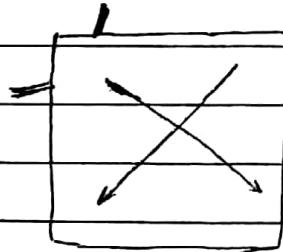
On Angiography \rightarrow Rt coronary artery fills.
- Lateral MI in infant.

Rx: Bypass Sx.

l-TGA \rightarrow Corrected TGA.

Normal Heart.

Atrio-Ventricular discordance.



[RA connected to LV
LA " " RV

No problem to baby.

KAWASAKI'S DS:

- M/c acquired heart disease in US, Japan, & Chandigarh.
- Fever > 5 days.
- Development of a limp.
- Erythematous macular exanthem over body.
- Ocular conjunctivitis.
- Dry & cracked lips.
- Red throat & cervical lymphadenopathy.
- Grade II/IV vibratory systolic ejection murmur at lower left sternal border.
- Predominant Neutrophils & ↑ platelet.
- M/c medium sized vasculitis.
- 20-25% causes develop aneurysm in future.

Q. Which of the following vasculitis not occur in adults?

A) Kawasaki's (85% are < 5 yrs)

B) Sjögren's Syndrome - Seen in adult females.

C) Giant cell arteritis

D) HSP

Peeling of palm & soles is classically seen in 2nd - 3rd wk of life.

Rare in B. Before 3 months of life.

DOC: IV Ig.

Δ: Fever > 5 days & any 4 of these 5

- ① Changes in extremities (eg: Erythema, edema, desquamation).
- ② B/l conjunctivitis (not assoc. with exudates).
- ③ Polymorphous rash (not vesicular).
- ④ Cervical lymphadenopathy.



⑤ Changes in lips & Oral cavity (eg: Pharyngeal erythema; dry/fissured or swollen lips, strawberry tongue).

Non-classical feature of Kawasaki:

Arthralgia; Arthritis;

Thrombocytosis; Urethritis;

aseptic meningitis (irritable)

M/C/C of death in Kawasaki —

Overall — Coronary artery aneurysm

Acute phase — Myocarditis.

Rx: 2v Ig 2g/kg in acute phase reduces risk 4-6%

Aspirin 100mg/kg/day x 2 weeks.

If Resistant to Ig: 10-15% cases

↳ Add steroid (Methylprednisolone).

↳ Repeat Ig

↳ TNF blockers → Infliximab; Etanercept.

↳ IL-1 inhibitor → ANAKINRA

Recurrence Rate → 1-2% cases.

Mx of aneurysm:

Small → 50% resolve over 1-2 yrs.

Aspirin 3-5mg/kg/day.

Medium to large → Add Warfarin

Sx (may be bypass)

HSP (Henoch-Schonlein Purpura):

M/c vasculitis (small vessel) Overall.

M/c leucocytoclastic vasculitis.

- Palpable non-thrombocytopenic purpura
- IgA deposition of vessels in dermis

Pathogenesis: Aberrant Galactosylation

Mesangio-proliferative disorder.

Small vessels → Skin

Arthritis

Arthralgia

GIT - mesenteric ischaemia

Kidneys - HSP Nephritis (40-50%).



84% develop in 4 weeks

91% " " 6 weeks

97% " " 6 months.

• Microscopic haematuria

• Proteinuria

• 1-2% RPGN (over days to week).

↳ On Biopsy crescent seen

↳ ∴ Crescentic GN



proliferation of parietal
epithelial cells.

Rx: i.v. Methylprednisolone.



Rheumatic Fever:

- M/c acquired heart ds in India / Developing country.
- Due to Group A β -hemolytic streptococci strains M-type 1, 3, 5, 6 & 18.
- Most frequent b/w 5-15 yrs.
- Latent period 3 weeks.
- Autoimmune: Molecular mimicry

Affects \leftarrow Myocardium
Muscle.

Modified Jones (2015):

- Era of Echocardiography \rightarrow Subclinical AR/MR.
- A/c to Risk area.

Low Risk - defined as having an ARF incidence
 < 2 per 100000 school-aged children.
 (usually 5-14 yrs old) per year or an
 all age prevalence of RHD of ≤ 1 per 1000
 population per year (Class 1B; level of
 evidence C).

Criteria = 2 major or 1 major + 2 minor
 + Essential criteria.

Recurrence = 2 major or 1 major + 2 minor or
 (OR)
 3 minor

Major Criteria:

Low risk populations:

- Carditis (clinical &/or subclinical)
- Arthritis (Polyarthritis only).
- Chorea
- Erythema marginatum
- Subcutaneous nodules.

Moderate to high-risk populations:

- Carditis (clinical &/or subclinical)
- Arthritis
 - Monoarthritis or polyarthritis
 - Polyarthralgia \oplus
- Chorea
- Erythema marginatum
- Subcutaneous nodules.

Minor Criteria:

Low risk populations:

- Polyarthralgia
- Fever ($\geq 38.5^{\circ}\text{C}$)
- $\text{ESR} \geq 60 \text{ mm in the first hour}$ &/or $\text{CRP} \geq 3.0 \text{ mg/dL}$
- Prolonged PR interval, after accounting for age variability.

Moderate to High risk populations:

- Monoarthralgia
- Fever $\geq 38.5^{\circ}\text{C}$
- $\text{ESR} \geq 30 \text{ mm in first hour}$ or $\text{CRP} > 3 \text{ mg/dL}$
- Prolonged PR interval - Also in MR & MS.



Rx:

- Aspirin 100mg/kg/day \times 12 wks.
- Prednisolone \times 12 wks.
 - 2mg/kg \times 3wks \rightarrow taper
 - Severe carditis or CHF.
- Crystalline Penicilline G \times 10 days
- IE prophylaxis.

Mitral valve \rightarrow M/c involved in RF

\hookrightarrow Recurrence cause Mitral stenosis.



Primary prevention



In India 2^o prevention



\hookrightarrow Penicillin G

Benzathine (i.m.)

- 600,000 IU for children wt \leq 60lb

1.2 million IU for children wt $>$ 60lb
every 4 wks.

or, Penicillin V (Oral)

\hookrightarrow 250 mg twice a day

or, Sulfadiazine or Sulfisoxazole (oral)

\hookrightarrow 0.5g once a day for pt. wt \leq 60lb

1.0g once a day for pt. wt $>$ 60 lb.

For pt. allergic to penicillin & Sulfa group.



Microlides are given orally.

CATEGORY	DURATION
- Rheumatic Fever \ominus out Carditis	5yr or until 21 yrs of age whichever is longer.
- RF \ominus carditis but \ominus residual heart ds (No valvular ds)	10yr or until 21 yrs of age, whichever is longer.
- RF \ominus carditis & Residual heart ds (persistent Valvular ds)	10 yrs or until 40 yrs of age whichever is longer. Sometimes lifelong prophylaxis.

Q

- Blood pressure = 86/600 mm Hg

- 4 yr, unconsciousness.

- HR = 180/min

- CFT = 4 sec

$\Delta = ?$ \rightarrow Compensated Shock.

Rx = 20 ml/kg 0.9% NaCl.

SHOCK = BP < 10th centile for age & sex.

Criteria for Shock:

Criteria for Hypotension by age.

Age	Systolic BP
Term neonates (0 to 28 days)	< 60 mm Hg
Infants (1-12 month)	< 70 mm Hg
Children (1-10 yrs)	< 70 + (age in yrs X 2)
Children > 10 yrs	< 90 mm Hg

Compensated shock: Rx - 20 ml/kg 0.9% NaCl

Repeat upto 60 ml/kg

\hookrightarrow CVP line (next step).

• low - fluid given.

- Normal → Cold epinephrin
- Warm: Nor-epinephrin.

Hypertension $BP > 95^{\text{th}} \text{ centile}$

- Essential ~10% ; increasing
- Secondary HTN

↳ Renal parenchymal - Reflux Nephropathy
VUR

↳ Renovascular

- Major → RAS; RVT

Minor → HUS

↳ Cardiac - post ductal coarctation (Tunne)

↳ Endocrinol - • Hyperthyroid

• Cushing

• Pheochromocytoma.

• CAH 11 beta/17 alpha

hydroxylase deficiency

End organ damage in HTN :

- Fundus
- Echocardiography concentric LVH.
- Any adolescent $BP > 130/80$
In children $> 10.4 \text{ yrs} \rightarrow 120/80 \rightarrow \text{HTN}$
- Urine protein.

Rx : HTN

- Life style modification.
- Pharmacological therapy - end organ; symptoms;
Severe $> 99^{\text{th}} \text{ centile}$.

↓
ACEi/ARBs.

ACEi C/I if GFR < 30 → causes hyperkalemia

↓
Rx → Amlodipine (ccb's)

Hypertensive Emergency:

- LVF \Rightarrow S₃; Gallop; Basal crepts.
- Seizures.

R:

Best: i.v. Nicardipine infusion

R: Sodium Nitroprusside i.v./
Esmolol/
Labetalol i.v.

#. Linezolid, Nitroprusside & Amphotericin B
should be covered.

Q. A 12yr old boy c seizure.

BP = 200/140.

Femoral pulses not palpable.

$\Delta = ?$

A) Takayasu aortoarteritis (R- Prednisolone).

B) Grand Mal Seizures.

C) Fibromuscular dysplasia (FMD)

D) Renal parenchymal defect.



Tachyarrhythmias

- Based on QRS.

~~wide QRS~~

Wide > 0.09 sec (VT/VF)

- Pulseless

- Stable

- Arrest.

Rx - lignocaine

Rx - Defibrillation $0.5 - 1$ J/kg

Amiodarone

Narrow

- Recurrent SVT

- HR > 220 infant; > 180 older

- P-wave absent; inverted

Vagal maneuvers

Stable

Rx - Adenosine (fastly given \pm saline flush)



as close to the heart
as possible.

CHF; Shock

Rx - Synchronized cardioversion

Respiratory

High & cold caused by Rhinovirus.

JMNCS (7L)

Age = 2m - 12 months = RR \approx 50 or more.

Age 12m - 60m = RR \approx 40 or more.

• Chest is Indrawing = Pneumonia.
or fast breathing

Rx :- Give oral Amoxicillin for 5 day.

Any general danger sign \Rightarrow Severe Pneumonia.
or Stridor in calm child { or very severe dz

R

\rightarrow Give 1st dose of an appropriate A/b.
 \rightarrow Refer to hospital.

Q. 18 m old child weighing 11.5 kg comes to PHC with resp. difficulty. O/E lethargic RR = 46/min.
no chest retraction = Next Step.

Ans (b) Prescribe oral antibiotic & Refer to higher centre.

\rightarrow Signs (< 2 months)

Next Step

- Convulsions or

Serious = Give i.v.

- Fast breathing (≥ 60)

Bacterial \Rightarrow Gentamycin.

- Severe Chest Indrawing.

Sug[®]

- 10 or more skin pustules.

↓
Refer to
higher centre

or a big boil or

If axillary temp 37.5°C

or above or less than 35.5°C

or lethargic / unconscious

- less than 10 unit.

- Umbilicus red or draining \rightarrow local (= Give oral)
- Plus) pus discharge from:
 - ear or < 10 skin pustules. \rightarrow **Septic**
 - ear or skin pustules. \rightarrow **Septic**

- Causes of Pneumonia Age wise in India & world
- Neonates = Grp. B Staphylococcus, *E. coli*.
 < 3 weeks.
- $30k - 3m$ = Respiratory Syncytial Virus (RSV)
S. pneumoniae, *H. influenzae*.
- $4m - 4yrs$ = RSV, *S. pneumoniae*, *H. influenzae*.
 $> 5yrs$ = *M. pneumoniae*, *S. pneumoniae*.
- *H. influenzae* $\xrightarrow{\text{dt}}$ Vaccination $\rightarrow \downarrow$ in Incidence.

Viral pneumonia

- Prodrome.
- Diffuse, Bl
- Not lobar pneumonia
- Interstitial infiltrate

T/E: ① Doc for RSV = Ribavirin.

② Influenza = Amantadine, Rimantadine

H1N1 = Influenza A \rightarrow Neuraminidase Inhibitors.

\hookrightarrow Oral Oseltamivir.

\hookrightarrow Inhaled Zanamivir.

Pandemic :-

Phase 1 - 3 Animals.

Phase 4 - May 27, 2009 (Mexico)

\hookrightarrow Human-human transmission at community

Phase 5 - Aug 5/09 OSA.

Human - Human transmission in one zone

Phase 6 = June 11, 2009, US & India.

→ Human - human transmission in two zones

H₁N₁ Virus :-

- SS RNA.

- Belongs to Orthomyxoviridae.

- Size = 80 - 200 nm.

Types :- A, B, C.

Surface Ag = H (Hemagglutinin)
N (Neuraminidase)

As the reassortment occurs in swine flu.
= Swine flu.

⇒ Antigenic shift & drift - No vaccines made

Symptoms :- Flu like.

Complications of H₁N₁ → in High risk groups.

⇒ Pneumonia (viral)

⇒ Bacterial Superinfection.

⇒ ARDS like features.

High Risk groups :-

Comorbidity

→ They were associated with some mortality

→ Pregnancy

→ Nephrotic syndrome, Chronic illness, post transplant

→ <1 yr., >65 yr.

Indications of oseltamivir

① Give it to all suspected cases of H₁N₁.

② Give it to all confirmed case.

ASis by throat swab & Nasopharyngeal.

Swab is sent for Real time PCR.

③ H₁N₁ influenza

Give it to all household contacts, occupational

adults

(Post exposure in doctors)

Dose = 75 mg (up to 5d)

Prophylactic dose = 75 mg D₁ → 7-10 days.

zamamavir.

The dose = Two 5mg Inhalations (10mg total)
twice / day x 5 days.

Prophylaxis dose = two x 5mg Inhal[®] - 07.

Q: H₁N₁ 10 wks pregnant -

→ Give oseltamivir [not teratogenic]

→ T₂

↳ only in rats

Q: Pt on Rifampicin Induces hepatic CYP450.

gets H₁N₁ sy[®]

→ What happens to dose of oseltamivir

Ans = Remains same → Renal ex[®]

Q: Dose in hemodialysis

= oseltamivir & dose is reduced

Pneumonia

- St. pneumonia = causes severe, lobar pneumonia.

Incub/ Rd = 1-3 days

(R) } Ceftriaxone, DOC.

→ Resistant variety = DOC → Vanco + Ceftriaxone

(Vaccines) :- > 2 yrs. children = PPV ~ 23.

< 2 yrs = PCV - 7/11 = conjugated.

6, 10, 14 weeks, 1/3 booster at 15-18 months.

IGAVI :- Global Alliance for Vaccines & Immun[®]

- founded by Bill Gates & Melinda Gates

- free vaccines in poor country

Staph. Aureus

- Max Mortality (10-30%)
- Air filled cavity (Pneumatocele)
- also seen in d/t

1) Klebsiella

2) Kerosene oil poisoning

→ Pneumatocele can Rupture & develop pneumothorax

- S. aureus = 1/3rd of sepsis in children.
(Plus in pleural cavity)

T/t = ICDT.

DOC for S. aureus = clavulanic.

Vancomycin for MRSA.

- H. influenza = Usually part of S. sepsis.
- can have otitis, meningitis.

Rx = ampicillin & chloramphenicol.

20-40% are resistant.

Rx: (Ceftriaxone) → DOC.

Atypical pneumonia

→ Rate - < 4 yrs = ~~20 yrs~~ > 5 yrs. !

- Symptoms - dry cough.

- Interstitial pneumonia

- Organism = Mycoplasma chlamyde.

Rx: - Macrolides

15 month

Motor → Walks alone, crawls up stairs

Adaptive → Makes tower of 3 cubes, makes a line with crayon, inserts raisins in bottle

Language -唐语; follows simple commands, may name a familiar object (eg. ball) responds to his/her name.

Social → Indicates some desires or needs by pointing, hugs parents.

18 month

Motor - Runs stiffly, sits on small chair, walks up stair with 1 hand held, explores drawers & waste baskets.

Adaptive → Makes tower of 4 cubes, imitates scribbling, imitates vertical strokes, dumps raisins from bottle.

Language - 10 words (average), names pictures, identifies 1 or more parts of body

Social → Feeds self, seeks help when in trouble, may complain when wet or soiled, kisses parent with pucker.

24 month

Motor → Runs well, walks up & down stairs, 1 step at a time, opens door, climbs on

furniture, jumps.

Adaptive → Makes tower of 7 cubes (6 at 21 months)
 scribbles in circular pattern, imitates
 H₃ stroke, folds paper once imitatively
Language → Puts 3 words together (subject, verb, object)

Social → Handles spoon well, often talk about
 immediate experiences, helps to uncross,
 listens to stories when shown pictures

30 months

Motor → Goes up stairs alternating feet

Adaptive → Makes tower of 9 cubes, makes V & horizontal strokes, but generally will not join them to make cross, imitates circular stroke, forming closed figure

Language → Refers to self by pronoun 'I'
 Knows full name

Social → Helps put things away, pretends in play

36 months

Motor → Rides tricycle, stands momentarily on 1 foot

Adaptive → Makes tower of 10 cubes, imitates construction of bridge of 3 cubes

copies circle, ~~imitates cross~~

Language - Knows age & sex, counts 3 objects
incorrectly, repeats 3 numbers or a sentence of 6 syllables, most of speech intelligible to strangers.

Social → Plays simple games (in parallel with other children) helps in dressing (unbuttons clothing & puts on shoes) washes hands.

48 months

Motor → Hops on 1 foot, throws ball overhead, uses scissors to cut out pictures, climbs well

Adaptive → Copies bridges from ~~parallel~~ model, imitates construction of gate of 5 cubes, copies cross & square, draws man with 2-4 parts besides head, identifies longer of 2 lines.

Language - Counts 4 pennies accurately, tells story

Social → Plays with several children, with beginning of social interaction & role-playing, goes to toilet alone

60 monthly.

Motor - Skips

Adaptive → Draws Δ from copy, names heavier of 2 weights

Language - Names 4 colours, repeats sentence of 10 syllables counts 10 pennies correctly

Social → Dresses & undresses, asks questions about meaning of words, engages in domestic role playing.



Fever & Stridor. Inspiratory sound in upper airway.

Croup / LTB

- 75% parainfluenza

→ Prodrome → Stridor

Barking Cough.

Subglottic narrowing

K/A = Stippled sign

On X-ray:

↳ Doc = formild = dexta = 0.6 mg/kg. Ceftriazone + Sulbactam mod & severe (ESBL)

Epiglottitis

In world - *S. pyogenes*.

S. pneumoniae

S. aureus

In India - *H. influenza*.

dat X-Ray → ^{Neck} Thumb Sign.

Rx → Airway [Emergency tracheostomy]

↳ Nebulisation & Racemic epinephrine.

↳ Westley Croup Score Criteria to dx it.

BRONCHIOLITIS:

- Inflammatory obstruction of smaller airway.

M/c organism - RSV (50%)

↳ Respiratory Syncytial Virus.

R/F:

- Males, Top fed.

① - Preterm; Chronic lung disease.

② - L → R shunt.

- Smoking mothers.

Airway resistance, $R = \frac{1}{r_4}$ → air trapping

Prodrome → Wheeze / ~~or~~ Ronchi.

X-Ray chest shows Hyper inflation.

Rx: Antibiotics has no role.

Humidified O₂.

For 2 High Risk group - Nebulised Ribavirin
Palivizumab

Long term - Persistent wheeze in infancy

↓
Reactive airway disease.

ASTHMA

Classification a/c to severity.

Step	Symptoms	Night symptoms	Peak expiratory flow rate
I.	Intermittent. <1 time a week; asymptomatic & (N) PEFR b/w attacks	≤ 2 times a month	≥ 80% predicted; Variability <20%
II.	Mild persistent. >1 time a week but <1 time a day	>2 times a month	≥ 80% predicted; Variability 20-30%
III.	Moderate persistent. Daily use β -agonists; daily attacks affect activity.	>1 times a month	>60% & <80% predicted; Variability >30%
IV	Severe persistent. Continuous limited frequent activity.	Frequent.	≤ 60% predicted; variability >30%

For intermittent → SOS β_2 -agonist

Mild persistent → +++ inhaled beclomethasone,
fluticasone, Budesonide.

Moderate persistent → +++ Salbutamol / Sustained release theophylline.

Severe persistent → ++ Oral low dose, long term, alternate day prednisolone.



2 yrs → Acute severe asthma



Oxygen; Nebulise salbutamol



Nebulised ipratropium bromide



i.v. hydrocortisone



s/c Terbutaline



Terbutaline infusion



50% $MgSO_4$ → aminophylline.

Foreign body:

$C_x R$ → Persistent inflation

Ball valve inflation.

R → Bronchoscopy & remove foreign body.

Recurrent pneumonia:

2 episodes of radiographic pneumonia in 1 year

(OR)

3 episodes in any time frame.

Persistent pneumonia:

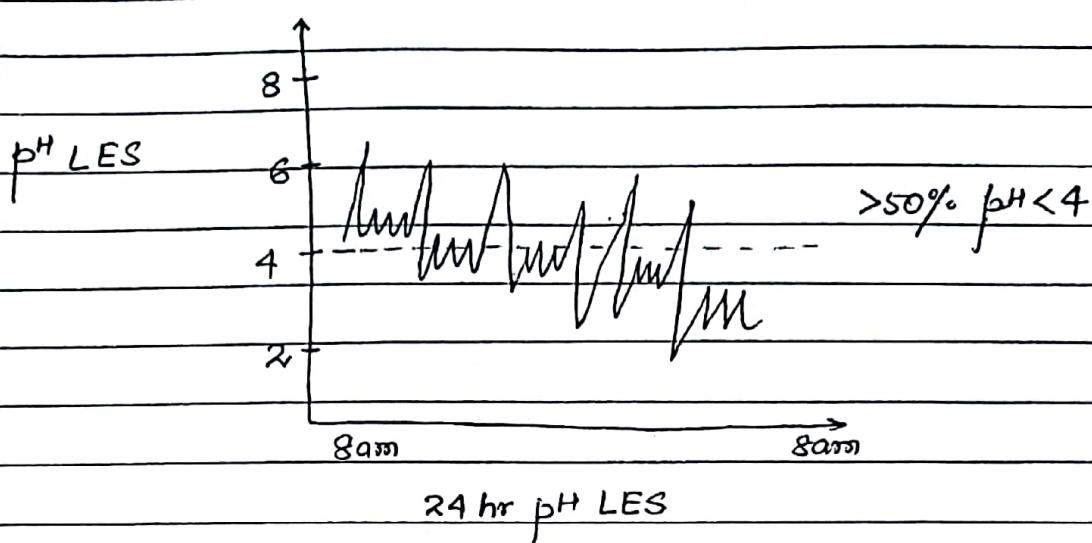
Persistence of symptoms & radiographic abnormalities for more than 1 month.

Recurrent pneumonia:

Cause → L → R shunt

• GERD dt aspiration.

Immunodeficiency.



M/c/c of Recurrent pneumonia -in U.S. = Cystic Fibrosis.

Cystic Fibrosis

- Incidence: 1 in 2500 in UK

Gene \rightarrow CFTR gene (7q 31.2 locus)



CAMP regulated chloride channel.

- In CF more Sodium goes to lumen \rightarrow to mucus than chloride.

- Autosomal Recessive disorder.

- Respiratory: Pneumonia (>5 yrs of life).

M/c organism ass. c cystic fibrosis - *Pseudomonas aeruginosa*.

(Mucoid $>$ Non Mucoid)

Rx:  Inhaled antibiotics

- Tobramycin
- Aztreonam
- Colistin
- Ciproflox

- Amikacin
- Levofloxacin

Respiratory : Pneumonia (>5 yrs)

- < 5 yrs - *S. aureus*; *H. influenzae*.
- 5-18 yrs - *S. aureus*; *Pseudomonas*
- > 18 yrs - *Pseudomonas*; *S. aureus*.

Acromonas; *Achromobacter*;
Burkholderia capsici

↳ specific; fatal

↳ Rx Mild - *Ceftriaxone*

↳ Severe - *Meropenem* +

Ceftriaxone / *Doxycycline*.

- Exocrine Pancreas - Steatorrhoea (< 5 yrs)
foul - bulky stools.

Rx - Steatorrhoea in CF

Lipase 1000 IU/kg supplement.

- Endocrine → 25% DM after 35 yrs.
- GIT → Meconium ileum.

Adolescents - Distal intestinal obstruction syndrome.

48hrs old baby has not passed meconium.

Ix → Lower GI contrast study

Diagnostic Therapeutic in meconium ileum.

- GIT: Diffuse pain abdomen
Colonic mucosal thickening.
Intussusceptions.

- Nasal polyps; Azoospermia

- Absent Vas: 1% infertile men; common in CF males.

Q. Which glands are not obstructed in CF?

- A) Cervix → Infertile
- B) Pancreas → Insufficiency.
- C) Sweat glands

→ causes "Trafficking Defect"

M/c mutation in CF = ΔF508 mutation (Class II mutation)
(Seen in 70% caucasians)
↓ (25-30% Indian)

M/c lethal genetic disorder in caucasians.

CFTR gene has 1800 mutations.

How many Nucleotide deleted in CF?

3 → TTT = Phenylalanine.

Rx: For Trafficking Defect

↓

DOC [LUMACAFTOR (Trafficking corrector drug)
+]

IVACAFTOR (Potentiator drug).

↓

opens CFTR Cl⁻ channel.

Lumacaftor - induce hepatic Cyp 450

↓

∴ New Corrector → TEZACAFTOR

↓

doesn't induce Cyp 450.



Q. Which of the following is a calcineurin inhibitor?

- A) Tacrolimus
- B) Sirolimus \rightarrow m-for inhibitor
- C) Everolimus
- D) Cyclosporine

Diagnostic Criteria of CF:

- ① Sweat Chloride $> 80 \text{ meq/L}$ on 2 occasions
[N $< 40 \text{ meq/L}$]
- OR, ② Two known CFTR mutations.
- OR, Best - ③ Diagnostic nasal electrode potential difference.

Newborn screening for CF

IRT test \rightarrow Immuno reactive Trypsinogen
Assay test.
(Sensitive test)

Q. Male, 10 month boy, Down Syndrome & Recurrent pneumonia

Filling defect \rightarrow In Ba-swallow

\hookrightarrow in middle of esophagus.

\hookrightarrow Aberrant right subclavian artery
vascular ring

\downarrow
Dysphagia lusoria.

Approach to a pt. c recurrent / persistent pneumonia:

History, physical exam, CXR

↓
— Rule out TB

Difficulty in
feeding: choking
during feeds.

Infection in other
parts of body

Associated
Malabsorption.
Pseudomemos

No clues

GER studies.

Immunoglobulin
CD4, CD8

in airway

Esophageal pH monitoring,
Barium Swallow.

NBT, HIV test.

Sweat Cl⁻ test

Direct laryngoscopy.

Mutalion Studies.

Iso tonic fluids:

- 0.9% NaCl — 154 meq/L Na & Cl.

- Ringer lactate = Plasma

- 130 meq/L Na

- 109 meq/L Cl

- 28 meq/L Lactate

- 4 meq/L K

- 3 meq/L Ca

Maintenance fluid:

Need?

- Insensible water loss.

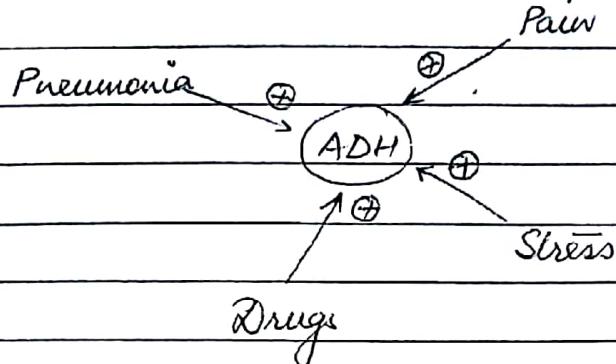
- Energy

- To prevent catabolism.



Type?

- Adult: 5% DNS
- Children: 5% Dextrose + N/2 or NS.



Amount?

A/C to Holiday & Segar

< 10 kg - 100 ml/kg/day.

10 kg - 1000 ml/day. (40 ml/hr).

11-20 kg → 1000 ml (for 10 kg) + 50 ml/kg additional kg above 10 kg.

> 20 kg → 1000 ml + 500 ml + 20 ml/kg additional above 20 kg.

Patau's Syndrome (Trisomy 13):

- Cleft lip & palate.

- Polydactyly

- Hypotelorism (Eye separated wide).

- Abnormal looks like Cyclops

↳ Holoprosencephaly.
(single eye)

↳ Fused frontal lobes + lateral ventricles.

↳ Aplasia cutis (problem of cleavage of skin).

- Rocker bottom

Edward Syndrome (Trisomy 18)

- Rocker bottom feet
- Overlapping of fingers.
- 2nd M/C Trisomy.
- Maternal age
- GIT anomaly are common
 - ↳ Atresia gut
 - ↳ Exomphalos
 - ↳ Malabsorption.

TTNB (Transient Tachypnoea of New Born): Wet lungs.

R/F :

- Term ; Cesarean Section
- Macrosomia
- Precipitous labour.
- Maternal Sedation

CxR - Prominent horizontal fissure.

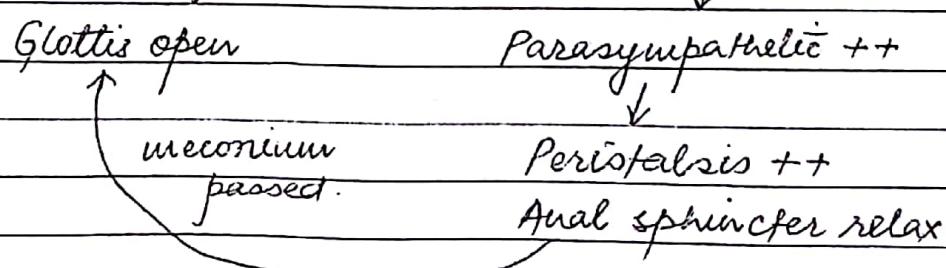
- It is a benign, self limiting condⁿ & resolves in 48-72 hrs.

FiO_2 requirement < 0.4

Never require mechanical ventilation.

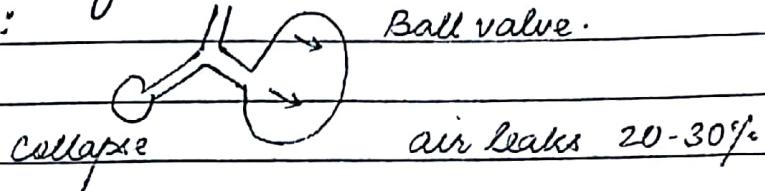
MSL (Meconium Stained Liquor):

- Marker of perinatal hypoxia (common in Post term)



Complications of Meconium:

Physical :



Ball valve.

Chemical : Irritant.

Biological \leftarrow impairs surfactant function (Surfactant _{given})
 Good culture media (Given Antibiotic)

Rx : Meconium stained liquor



Born

Vigorous \leftarrow Zone
 HR $> 100/\text{min}$
 Resp effort.

YES

Transfer to mother

Liap or,
 HR > 100 or,
 Apnea.

NO
 \rightarrow
 PPV $\times 100\%$ O₂.

GROWTH

1-4 months : Weight gain 30gm/day

5-8 month : wt. gain @ 20gm/day

9-12 month : wt. gain @ 10gm/day.

Weight multiples:

wt. \times 2 = 5 months

\times 3 = 1 yr

\times 4 = 2 yrs

\times 5 = 3 yrs

\times 6 = 5 yrs

\times 7 = 7 yrs

\times 10 = 10 yrs.

Length:

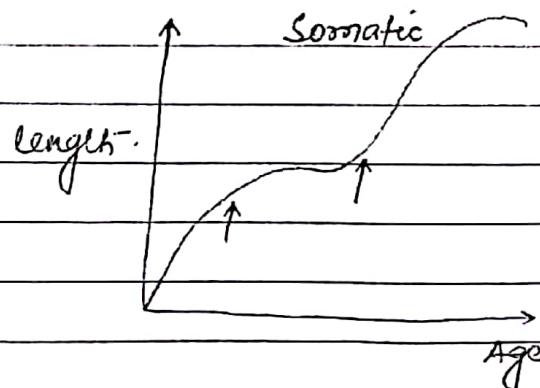
At birth - 50cm] Growth Velocity $\rightarrow +25$ (first year)

1 yr - 75cm

2 yrs - 90cm $\rightarrow +15$ \rightarrow second year.

$4\frac{1}{2}$ yrs - 100cm

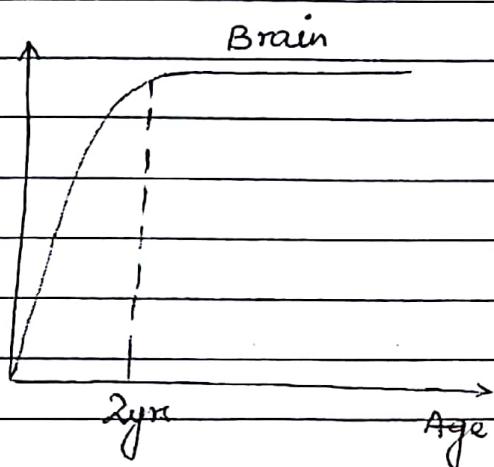
Add 6cm/year till puberty.



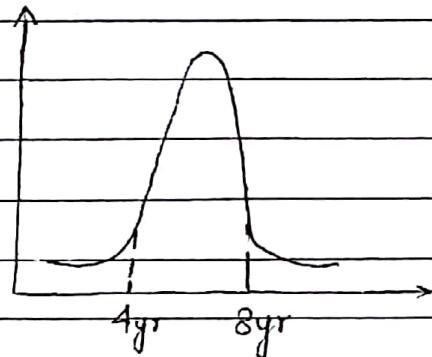
Q. In school going children; the avg height velocity is
 $\rightarrow 5-8$ cm

Head circumference:

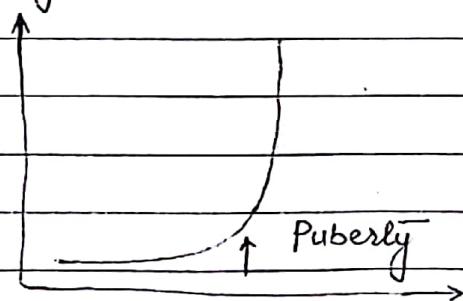
- 35cm at birth
- 3 months → 40cm
- 12 months → 45cm
- 2 yrs → 48cm (90% of brain grows)
- 12 yrs → 52cm.



Lymphoid growth:



Gonad growth:



Adolescent — 10 to 19 yrs.

Tanner's Sexual Maturity Rating:

SMR Stage I - V

Stage I — No character

Stage V — Completely developed.

Menarche → SMR - IV

Sequence of puberty:

Girls:

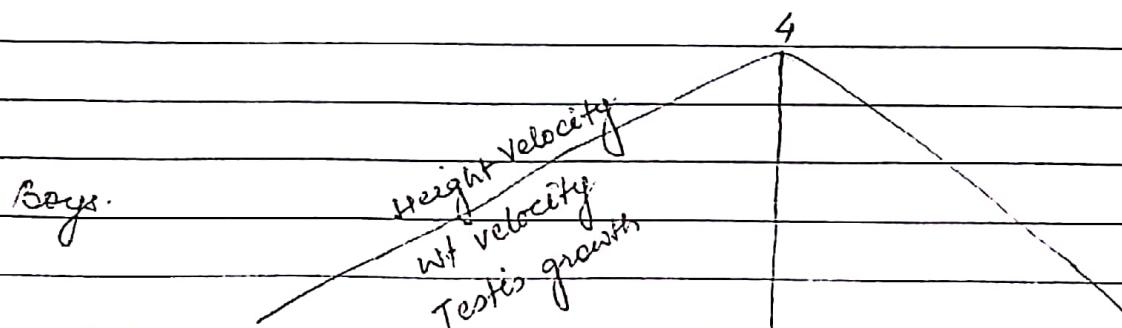
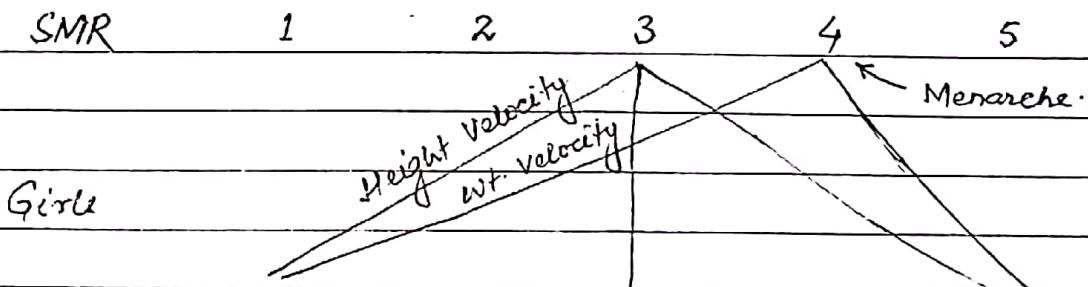
Growth Spurt → Thelarche, Pubarche, Menarche.

Boys:

→ Testis — Penis — Pubic hair, Axillary hair.

Girls: BP (Breast — Pubic hair)

Boys: GP (Genitalia — Pubic hair).



Most rapid ↑ in height → Phase 3 in girls & Phase 4 in boys.

Puberty - comes by pulsatile release of GnRH.

Precocious Puberty:

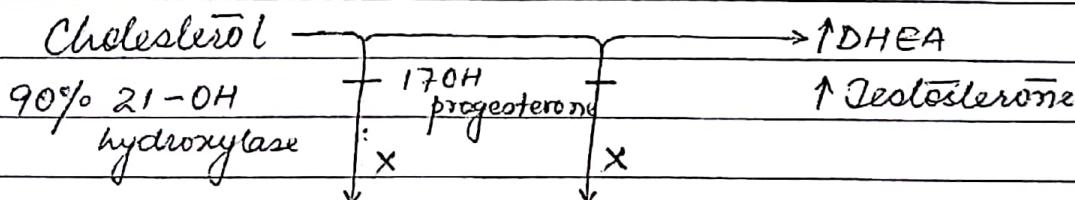
- Gonadotropin dependent / Central
- Females - Idiopathic.
- Males - Organic
 - (Hypothalamic Hamartoma, Craniopharyngioma, Hydrocephalus, TBM).

CAH (Congenital Adrenal Hyperplasia):

M/c/c of female hermaphrodite.

Karyotype - XX

But looks like Male.



Cortisol Aldosterone

glucose ↓ Na^+ ↓

Salt-Wasting Crisis (70%)

$\uparrow \text{K}^+$

Shock

- Cause of precocious puberty in boys.

Genitalia → Ambiguous

Labia is pigmented d/t ACTH.

Virilization

Hypertrophy of clitoris

- Penoscrotal hypospadias
 + Empty Scrotum.

Due to deficiency of 21-OH hydroxylase lead to accumulation of 17-OH progesterone.

♀ Level of 17-OH progesterone in CAH

A) < 150

B) $150 - 300$

C) $300 - 500$

D) $> 600 (> 3500)$

CAH associated with Premature epiphyseal closure.



Short Stature.

Rx: CAH

- Supplement of Hydrocortisone & Fludrocortisone



Glucocorticoids

Mineralocorticoids.

$15-20 \text{ mg/m}^2/\text{day}$

0.15 mg/day .

Thrice daily.

- Girls require Sx

- Clitoroplasty

- Reconstruction Sx.

- Never get married.

Cholesterol

11β -OH

deficiency

11-deoxycortisol

Cortisol

DOC_A

potent

mineralocorticoid

↓

Aldosterone

HTN

Hypokalemia metabolic
 alkalosis.

Q. 5 yrs old boy has precocious puberty. BP = 130/80
Estimation of cGH helps diagnosis?

- A) 17-OH progesterone
- B) 11-deoxy cortisol
- C) Aldosterone
- D) DOCA

Deficiency of 17 α -Hydroxylase \rightarrow \uparrow Aldosterone

\downarrow
HTN, Hypokalemia,
metabolic alkalosis.

\rightarrow \times Cortisol
 \downarrow glucose.

\rightarrow \times Testosterone

\downarrow

Male look like ♀.

(Mineralocorticoid) M

T (Testosterone).

α

\uparrow

\uparrow

\uparrow

\uparrow

\uparrow

3 β Hydroxysteroid dehydrogenase deficiency (3 β HSD def.)

\downarrow
Causes ambiguous genitalia in both sexes.

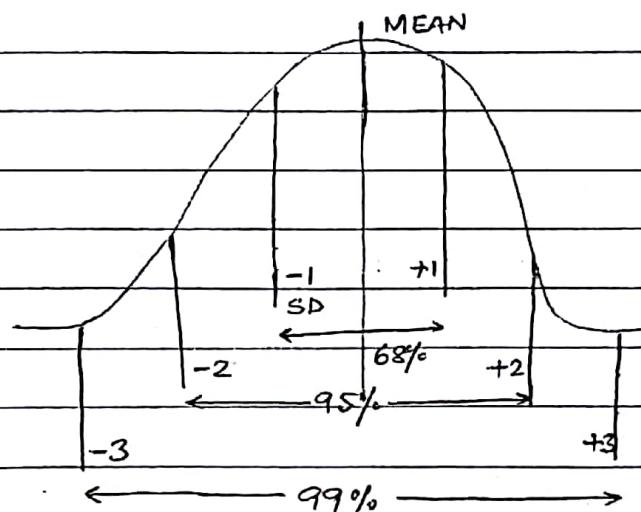
Q. Mother; previous child CAH

Next pregnancy - To prevent female virilization
 of foetus

Dexamethasone

(20mcg/kg pre pregnancy wt.)

Inhibits ACTH.



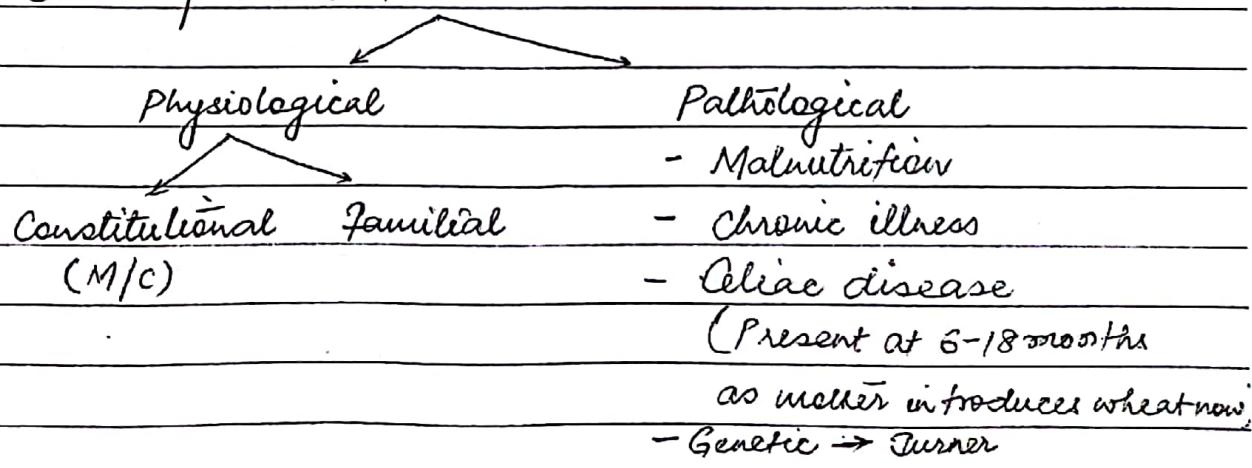
Short height > -2 SD below mean or $<$ 3rd percentile

Mid parental height = Adult predicted height

Boys = average parents height + 6.5cm

Girls = " " - 6.5cm.

Causes of Short Stature:



Rx: Celiac ds

- Restrict wheat, Rye & Barley for lifetime.
- Gluten-free diet.
- Also avoid oats.



- Endocrinopathy → Hypothyroidism
Cushing
Cushing
Growth Hormone deficiency.

Growth Hormone deficiency:

- Birth weight & length normal.
- Lag at 1-2 yrs of life.
- Bone age delayed.
- Doll face
- Micro penis] Panhypopituitarism
- Short stature

#. Micro penis (\downarrow LH, \downarrow FSH)

\downarrow ACTH → Hypoglycemia

- Hoarseness of voice.

Investigations: GH level \Rightarrow IGF, BP3

Growth Hormone stimulation test - Best

↳ Basal level GH

↳ Stimulated level - 2 stimuli

(clonidine; insulin; L-arginine).

Rx - Recombinant GH (US, FDA)

Indication: rGH

- GH deficiency
- Turner Syndrome
- Prader-Willi
- Chronic kidney ds.
- SGA height - 2-2.5 SD below mean

When to stop GH?

- Height reaches 50th centile
- Epiphysis fuses.
- Pseudotumor cerebri
- Slipped capital femoral epiphysis.

Disproportionate short stature:

US/LS ↑ → Achondroplasia, Rickets;

Hypothyroidism.

US/LS ↓ → TB spine, Mucopolysaccharoidosis IV

(Morgagni's ds)

Physiological short stature:

	CONSTITUTIONAL (M/c)	FAMILIAL
Birth length	• (N)	IUGR
Lag	6-12 months	
Growth velocity	(N)	Less
Final Height	(N) / Sub (N)	Less
Puberty	Delayed	(N) \neq
Bone age	Delayed.	(N) \neq



Developmental milestones:

Neck holding → @ 3 month.

Gross motor milestones :

3 months - Head holding; Neck holding.

5 months - Sitting \pm support.

4-6 months - Prone to supine, supine to prone
(Roll in bed).

8 months - Sitting \pm out support.

9 months - Crawling

10 " - Creeping; stand \pm support

12 " - Standing \pm out support; walking \pm out support.

2 yr - Walk up stairs \pm two feet at each step.

3 yr - Upstairs \pm one foot at each step, rides tricycle.

4 yr - Hop on one foot

5 yr - Skips on two foot

Fine motor :

12 wks \rightarrow Moro's reflex disappears

Grasp reflex disappears

4 months - Goes for objects

5 months - Bidextrous grasp.

6-7 " - Transfer object, palmar grasp.

9 " - Pincher grasp; mature meat.

13 months - Casting.

15 " - Self feed \pm a spoon

18 " - Self feed \pm a cup.

24 " - turns pages of a book one at time.

Social Milestone:

- 2 months - Social smile
- 3 " - Recognizes mother.
- 6-7 " - Smiles at mirror image.
- 9 months - Waves bye-bye.
- 6 months - Stranger anxiety.
- 2 yrs - Dry by day
- 3 yrs - Dry by night
- Dress/ undress himself (Supervision)
 ↓
 Out supervision - 5 yrs.

18 months - Separation anxiety/ Clinginess/ Reproachment.

Language Milestones:

1 month → Head turns to sound

3 months → Cooing

6 months → Babbles; Monosyllables (ma, ba)

9 months → Bisyllables (mama; ba-ba)

1 yr → 2 words in meaning

18 months → Vocabulary of ten words.

2 yrs → Simple sentences in 2 words; Phrase.

3 yrs → tells age & sex; uses pronouns,

handedness; identify colours.

4 yrs → tells story

5 yrs → Knows colour

15 months: 4-5 words

2 yrs → 50-100 words

When can a child understand death =

< 3 yrs - No idea

3-9 yrs - Idea

> 9 yrs - Entity; Irreversible; himself

Object permanence / Constancy - 9 months.

Cross a busy road - 10-12 yrs.

Tie shoe lace - 5 yrs.

Bladder control - 85% by 5 yrs.

Nocturnal enuresis - > 5 yrs.

↳ more than 2/week x 3 months.

Q. 14 yrs old child c Nocturnal enuresis

Rx - • Behavioural - +ve reinforcement.*

• Alarm therapy (Best)

↳ 85% relapse free rates.

• Drugs → Desmopressin

Encopresis : > 4 yrs

Cause → Chronic constipation.



CNS

MICROCEPHALY:

→ HC > -3 SD below mean

Causes: GENETIC

- Trisomy 13, 18, 21
- Cri-du-chat syndrome

ACQUIRED

- Baby: HIE; Hypoglycaemia, PKU, Meningitis, encephalitis.
- Mother → TORCH, ^{lalanine} Hyperphenylalanine $> 6 \text{ mg/dl}$
Alcohol, DM, Radiation.

MACROCEPHALY

HC > 2 SD above mean

Q Neurodegenerative disorders:

GREY MATTER:

- Normal at birth.
- Regression milestones.
 - ↳ Disappear as they come orderly.
- Deaf, blind, seizures.
- Anemia, Hepatosplenomegaly.
- Cherry red spot macula.

Microcephaly

Macrocephaly

- GM1 gangliosidosis.
- GM2 gangliosidosis
(Tay Sachs' ds)
- Gaucher's ds
(β -gluco
- Sandhoff
- Niemann Pick's ds

Huge
spleen

(

Tay Sachs ds -

- AR
- β /t deficiency of β -hexose aminidase A.
- 1/25 askenazi jew are carrier.
- 6 months exaggerated startle reflex.
- Cherry red spot macule
- Organomegaly not seen.

Organomegaly + Tay Sachs = Sandhoff



Def. of β -hexose aminidase A & B.

WHITE MATTER DISORDERS:

- All the tracts are white matter.
- Frequent fall, incoordination.
- Upper motor neuron sign.

Microcephaly / N

- Krabbe's
- XLR adrenoleukodystrophy.

↓
degeneration starts from
parieto-occipetal area.

△ \rightarrow ↑ VLCFA levels

Rx \rightarrow Early bone marrow Tx.

Lorenzo oil

- Metachromatic

leukodystrophy

Macrocephaly

Canavan ds.

Alexander ds.

→ MRI \rightarrow Diffuse white matter

thickening.

→ MRI \rightarrow Degeneration starts from
frontal periventricular area.



Hydrocephalus:

- Enlarged ventricles \rightarrow or \leftarrow out \uparrow in ICT.

CSF production:

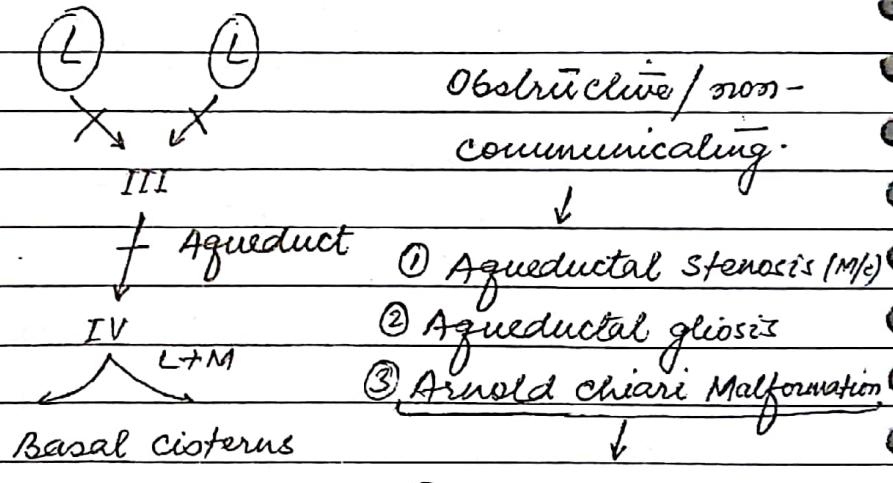
- Choroid plexus (75%) \rightarrow lateral, III & IV ventricles.
- Extrachoroidal (25%) \rightarrow Capillary endothelium in brain parenchyma.

Rate of CSF production \rightarrow 20 ml/hr.

CSF volume in infants = 50 ml

adults = 150 ml.

CSF flow:

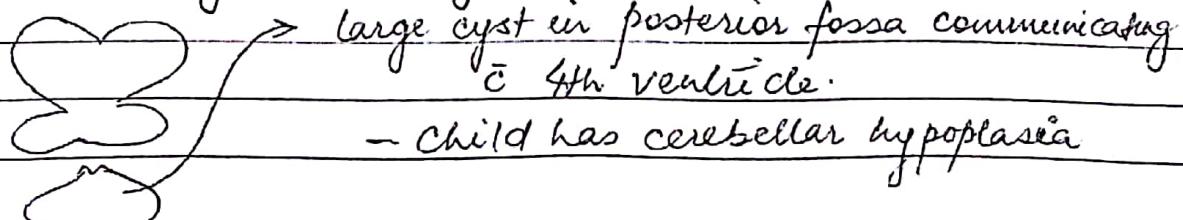


Arnold - Chiari Type I - Adolescent/Adult

Type II - Newborn/ \leftarrow

lumbosacral myelomeningocele

④ Dandy - Walker Syndrome:



⑤ Vein of Galen malformation:

- M/C arteriovenous malformation in brain
- Sinus Venosus ASD.
- Obstructs aqueduct.
- Midline mass in dilated lateral ventricle.

Non-obstructive / communicating:

- Basal exudates

↳ TB

↳ Cryptococcal meningitis.

IOC for congenital Hydrocephalus — MRI.

Rx: Drugs → Furosemide

Aclazolamide.

VP shunts → anastomose ~~to~~ ventricles to peritoneum.

↳ Gross hydrocephalus

↳ Deviated parenchyma.

↳ Complication — Blockage

Infection (Coagulase
-ve staph).

Febrile Seizures:

- M/c seizure during childhood.
- Between 6 months - 5 yrs.

R/F for recurrence:

Major:

Age < 1 yr

Duration of fever < 24 hr.

Fever 38-39°C (100.4-102.2°F)

Minor:

Family H/o Febrile seizures.

Family H/o epilepsy.

Complex febrile seizures.

Daycare

Male gender.

Lower Serum Na⁺ at time of presentation.

Having no risk factors carries a recurrence risk of approx 12%

if 1 R/F → 25-50%

2 " → 50-59%

≥ 3 " → 73-100%.

R/F	Risk for Subsequent Epilepsy
Simple febrile Seizure	1%
Recurrent "	4%
Complex " "	6%
(>15 min duration or recurrent > in 24 hrs)	
Fever < 1 before febrile seizure	11%
Family H/o epilepsy	18%
Neurodevelopmental abnormality (Mental Retardation)	33%

Complex febrile seizure complication:

- Prolonged febrile seizures.
MRI → Mesial temporal sclerosis

Temporal lobe + Hippocampal lobe.

Epilepsy in children:

Partial:

- Simple
- Complex - aura; automatisms

Rx - Oxcarbamazepine &
Carbamazepine.

Ring Enhancing lesions.

Neurocysticercosis

- Solitary
- SCOLEX

Rx - Albendazole (DOC)

↓

Before giving Albendazole

3-5 days of steroids given.

Tuberculoma.

- Large $> 20\text{mm}$
- Multiple
- Irregular margin
- Perilesional edema

- Midline shift.

Generalised epilepsy:

- Tonic
- Clonic
- GTCS [aura - GTCS → postictal phase]

↓

Drowsy, unconscious,
frothing, tongue bite
uprolling, incontinence.

- Atonic
- Myotonic

Rx - Sodium Valproate.

↓
In < 2 yrs → It is hepatotoxic.

Absence Seizure:

- Blank stare < 30 sec.
- No aura/post ictal phase.
- Hyperventilation provokes.

Rx : Ethosuximide (DOC)
Valproate

Atypical Absence seizure:

- Myoclonic component

Rx - Valproate.

JME (Juvenile Myoclonic epilepsy):

- 12-18 yrs.
- Myoclonic jerks morning.
- Drops things.
- Gene by gene
- Family History.
- GTCS seizures 90%
- 1/3 Absence seizures.

EEG of JME → Generalised 4-6 Hz spike
+ photic stimulation.

Rx - Valproate (lifelong) → Excellent.

Infantile spasms / Salaam / West

- Flexor contractions of Head, trunk & extremities.

- 4-8 months

- EEG → HYP SARRHYTHMIA

↳ Generalised chaotic high volume slow wave.

- Idiopathic / Cryptogenic → Good.

- Secondary → HIE, Structural malformation, Down's Syndrome, Tuberous sclerosis.

Rx: — ACTH → inhibit CRH. (DOC).

Vigabatrin → In Tuberous Sclerosis

STATUS EPILEPTICUS:

- Convulsion > 30 min

(OK)

Continuous b/w no regain of consciousness.

Vulnerable to hypoxia — Hippocampus, amygdala, Thalamus, subcortical areas.

Rx: — i.v. Lorazepam (DOC) — longer t_{1/2}
 0.05 mg/kg

- Midazolam + Phenytoin (20 mg/kg)

Repeat 10 mg/kg — 10 mg/kg

Phenobarbitone 20 mg/kg — 10 mg/kg

i.v. Valproate 20-30 mg/kg

Midazolam infusion 2-20 mcg/kg/min.

i.v. ~~Levetiracetam~~ Levetiracetam 20 mg/kg.

GA propofol; Thiopentone.



M/c/c of Status epilepticus → Febrile Seizures.

↓
Rx → per rectal diazepam /
Buccal midazolam

Prevention of Febrile Seizures:

- No need

- Risk of Recurrence / concerned parents



Intermittent prophylaxis Oral CLOBAZAM / DIAZEPAM

↳ New BZD

for 48-72 hrs of fever.

MENINGITIS:

Cause:

	India	World
< 2 months	Klebsiella E. coli	Gr. B / D Streptococci E. coli
2 months - 3 yrs	H. influenzae type B	S. pneumoniae.
> 3 yrs	S. pneumoniae	S. pneumoniae Nisseria

Acute Bacterial meningitis:

- 95% cases occur b/w 1 month to 5 yrs.
- Defect of complement system C₅-C₈ & properdin system
 - meningococcal infection
- T-lymphocyte defects (eg - AIDS/chemotherapy)
 - Listeria monocytogene / cryptococcus.

- congenital/acquired defects across mucocutaneous barrier → Pneumococci d/t cribiform plate.
- Lumbosacral meningocele & dermal sinus - staph. & enteric bacteria.
- Penetrating CNS trauma / CSF shunt infection
 - coagulase -ve staph.

Recurrent meningitis in CSF leak pt. M/c d/t pneumococcus.

Autosplenectomy (Sickle cell dysfunction/asplenia)
→ Pneumococcal infection }
H. influenzae } capsulated.
N. meningitidis

Splenectomy vaccination time - 2 wks before.

Rx: Ceftriaxone (DOC)

DOC in Resistant pneumococci = Vancomycin

+ Ceftriaxone.

M/c Neurological sequel of meningitis:

• SNHL via aqueduct cochlear.

• Can we prevent SNHL?

- Dexamethasone

- 0.15 mg/kg

- 30-60 mins before antibiotics.

Post exposure prophylaxis to contacts & doctors -

H. influenzae & N. meningitidis → i.m. Single dose ceftriaxone.

- Rifampicin X 2 days

Doctors → Fluoroquinolones.



Q. 3 yrs old diagnosed to have HiB meningitis. Tx done before discharge → BERA.
(Brainstem evoked response audiometry)

ENCEPHALITIS:

M/c/c = Enteroviruses (80% cases).

M/c sporadic = HSV-1

M/c aseptic meningitis in unimmunized children
— mumps.

Q. Child c fever & coma

Focal seizures.

CSF: Hemorrhagic

CT: Temporal hypodense; MRI → Hyperintense
— HSV-1 infection.

Localised temporal spike → HSV encephalitis.



DOC: i.v. Acyclovir.

Mortality rate of untreated herpes = 70%.

AFP (Acute Flaccid paralysis):

- Acute onset < 6 weeks

- < 15 yrs

- Rule out pseudoparalysis

↳ Septic arthritis

Osteomyelitis

— Scurvy; early syphilis

Hypokalemia → Hypotonia.

- Asymmetrical AFP:

• Paralytic polio.

• Traumatic neuritis \rightarrow i.m. injection (a/t)

- Symmetrical AFP :

• Transverse myelitis

\hookrightarrow Herpes, varicella, mycoplasma

Level \rightarrow Thoracic area.

Rx: high dose i.v. Methyl prednisolone.

• Guillain Barre Syndrome (ATDP)

\hookrightarrow Demyelinating.

- Diarrhea (By Campylobacter jejuni).

- Weakness occurs after 10 days.

- Areflexia (DTR absent)

- Symmetric

- Ascending \rightarrow diaphragm involved.

- Plateaus \rightarrow 4 wks.

- Sensory & autonomic changes.

- Also a/t Mycoplasma; salmonella;
S. pneumoniae.

CSF	1st wk	2nd wk
Cells	10/ulpf	10
Proteins	50 mg/dl	500

\rightarrow Abnormal cytological dissociation

- B/L symmetric demyelinating illness.

IOC for calcification: CT scan.

Rx: i.v. Ig (2g/kg)

\downarrow fails

Plasmapheresis.



IgG indications:

- ↳ Kawasaki
- ↳ AIDP (Acute Inflammatory Demyelinating Polyradiculoneuropathy)
- ↳ Hypogammaglobulinemia
- ↳ Rh isoimmunisation.

General Pediatrics:

- Diarrhea
- ORS
- Zinc
- Malnutrition:

ORS:

Residual	Mmol/L	Old (WHO-ORS)	New / Universal
45	Sodium	90 (Cholera stool loss)	75 (Rotavirus 50-70)
40	Potassium	20	20
125	Glucose	111	75 (for facilitate diffusion of Na ⁺ in cell)
70	Chloride	80	65
7	Citrate	10	10
+Mn, Zn, Cu		311	245
300			low osmolarity ORS.

Citrate improves the shelf life of ORS.

Resomal → Rehydration soln for malnutrition.

Hyperkalemia Hypernatremia.

Substrate concentration of components of ORS soln:

NaCl — 2.6 gms

KCl — 1.5 gms

Trisodium citrate — 2.9 gms

Glucose, anhydrous — 13.5 gms

WHO dehydration:

- No
- Some → Skin pinch slowly; thirsty
- Severe → skin pinch very slowly; lethargic; oliguria.

Rx: No dehydration:

- Replace ongoing losses.
- 5-10 ml/kg.

Some dehydration:

- ORS 75 ml/kg over 4 hrs.

Severe dehydration:

- i.v. Ringer lactate 100 ml/kg.

	30 ml/kg	70 ml/kg
>1 yr	0.5 hr	2.5 hrs
<1 yr	1 hr	5 hrs.

WHO Zn in Acute Diarrhea:

- 2x RDA for 2 weeks.

Zn dose	RDA	Diarrhea
>6 months	10 mg	20 mg.
<6 months	5 mg	10 mg

Aerodermatitis enteropathica (periorificial dermatitis)

- d/t Zn deficiency.
- Nutritional
- Genetic (AR) → Intestinal Zn transporter defect.
- Low Zn levels.

- Improve on Zn supplement.

Rx - Zn (3mg/kg) — elemental Zn.

Malnutrition:

	Marasmus	Kwashiorkar
Muscle Wasting	++	++
Edema		++
Hepatomegaly		++
Low albumin		++
Pigment		++
Aptite	Voracious	Poor
Sensorium	Alert	lethargy.

Weight/Age criteria (Indian Academy of Pediatrics)

- Normal $> 80\%$ reference.
- Grade I $\rightarrow 71 - 80\%$
- Grade II $\rightarrow 61 - 70\%$
- Grade III $\rightarrow 51 - 60\%$
- Grade IV $\rightarrow \leq 50\%$

for Edema \Rightarrow Add K

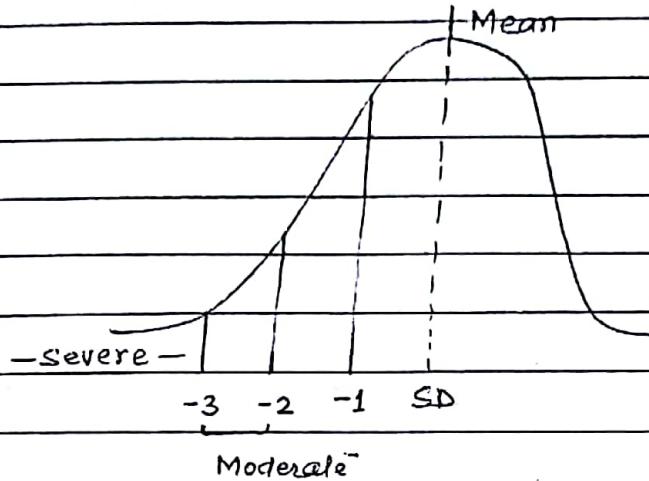
\hookrightarrow Bad marker.

WHO

	Moderate	Severe
Weight/Height	71 - 79%	≤ 70
Acute (wasting)		
(N) $> 80\%$ reference		
Height/age		
Chronic (stunting)	86 - 89%	$< 85\%$
(N) $> 90\%$ reference		

Symmetrical Edema

+++



Severe Acute Malnutrition:

Among children - 6-59 months of age.

Any of the following:

- ① Weight for height below -3 standard deviation (SD or Z scores) of the median WHO growth references.
- ② Visible severe wasting.
- ③ Presence of bimodal edema.
- ④ Mid-arm circumference below 11.5 cm.

Below 6 months → Mid-arm circumference can't be used.

Criteria for admission - If child fails appetite test.

Criteria for passing Appetite test.

Body wt. (kg)	Min ^{mm} amount of RUTF (Ready to use therapeutic factor) to be consumed for passing Appetite test (ml or grams)
< 4 kg	15 mL
4-6.9	25 mL

7-9.9 35 mL
10-14.9 50 mL

Mix:

Complications of SAM

1st & day $H = \text{Hypothermia} (< 95.5\text{F} \text{ OR } < 35.5^\circ\text{C})$

$\vec{c} \in \text{in 1st wk} \leftarrow I = \text{Infections (TB, Malaria, UTI)}$

EL = Electrolyte (Hypokalemia & Hypernatremia)

DE = Dehydration

D = Deficiency of Vitamins & minerals.

PHASE

STABILIZATION

REHABILITATION

Step	Day 1-2	Days 3-7	Weeks 2-6
1. Hypoglycemia	→		
2. Hypothermia	→		
3. Dehydration	→		
4. Electrolytes	→		
5. Infection	→		
6. Micronutrients	No iron	C 2300	→
7. Cautious feeding	→		
8. Catch up growth		→	
9. Sensory stimulation	→		
10. Prepare for follow up		→	

Feeding Rehabilitation: cautious feeding

- F-75 containing 75 kcal/100ml & 0.9 g protein/100ml.

B = Begin feeds

E = Energy dense feeds.

S = Stimulation

T = Tender Love & Care

Days	Frequency	Vol/kg/feed	Vol/kg/day
1-2	2 hrly	11 ml	130 ml
3-5	3 hrly	16 ml	130 ml
6-7	4 hrly	22 ml	130 ml

Energy dense feeds:

After the transition give:

- Frequent feeds (at least 4-hrly) of unlimited amounts of a catch-up formula.
- 150 - 220 kcal/kg/day.
- 4-6 gm protein/kg/day.

Criteria for discharge:

- Weight for height $> 80\%$ of Reference standard.
- Edema should be absent for 2 wks.
- MAC > 12.5 cm.
- Weight gain > 5 g/kg/day \times 3 days.
- Appetite is good.
- Complete antibiotics.
- Care taker should have learnt, motivated.

Weight for age $<$ Acute (wasting)
 Chronic (stunting)

Age independent criteria - MAC (> 12.5 cm)

b/w 1 to 5 yrs.

Age independent index - Kanawati & McLaren index

Rao & Singh index

Dugdale index.

Quac stick index

Telliffe ratio



$$\# \text{ Osmolality} = 2 \times [\text{Na}] + [\text{Glucose}] / 18 + [\text{BUN}] / 2.8.$$

Pediatric Nephrology:

- Development
- Oliguria; polyuria
- Hematuria
- AKI
- CKD
- Nephrotic, Nephritic
- UTI

Topic

Development:

GFR:

- Newborn $\rightarrow 15 - 20 \text{ m}\text{l/min} / 1.73 \text{ m}^2$
- 3 months $\rightarrow 2/3^{\text{rd}}$ adult
- Like adults $\rightarrow 2 \text{ yrs of life.}$

- Tubular concⁿ:

- Adult morning urine osmolality $> 800 \text{ mosm/kg.}$
- like adult 1 yr.

- Nephrogenesis complete @ 36 wk of gestation.

- Barker's hypothesis:

preterm & IUGR \rightarrow hypertension in 2nd to 3rd decade
 \hookrightarrow Bcoz they have less no. of nephron.

- M/c asymptomatic abd. mass in 1-5 yrs - Wilms tumour

- M/c abd. mass in newborn - Multicystic

dysplastic kidney.

now-functional mass \rightarrow bunch of grapes

- 80% U/L.

OLIGURIA:

- U.O. $< 1 \text{ ml/kg/hr}$
- Common in AKI & Acute GN.

Causes of Non oliguric AKI — Aminoglycosides

- Neonatal renal failure.
- Resolving ATN (polyuria)

POLYURIA:

- U.O. $> 4-5 \text{ ml/kg/hr}$. [Polydipsia; Polyuria]

Approach —

- Blood glucose [RBS $> 200 \text{ mg/dl}$
or FBS $> 126 \text{ mg/dl}$]

↳ DM

- Venous Blood Gas.

(i) Hypokalemic hypochloric metabolic alkalosis



Bartter Syndrome; Gitelman Syndrome.

Bartter Syndrome

- Severe; Early.
- Infancy
- Antenatal \rightarrow Polyhydramnios
- Hypercalcemia

Gitelman Syndrome

- Mild; Well preserved.
- Older child
- HypoMg
- Hypocalciuria.

(ii) Hyper cl, hyper k, Normal anion gap
metabolic acidosis



Renal tubular acidosis.

(iii) Normal \leftarrow Psychogenic \rightarrow water deprivation

DI

\rightarrow in psychogenic \rightarrow
urine osmolality doubles

DI \rightarrow Central / XLR Nephrogenic

- On Vasopressin challenge, urine osmolality doubles by 100% in central. Hence differentiated from XLR Nephrogenic.

Q 4yr; Polyuria & Polydipsia

Venous blood gas normal

Urine Osmolality

- Baseline 50 mosm/kg.
- On water deprivation = 60 mosm/kg
- On Vasopressin challenge = 70 mosm/kg.

Δ = XLR Nephrogenic DI

DOC = Thiazides.

Best method of GFR estimation = Insulin clearance.

Schwartz eGFR formula:

$$= \frac{K \times \text{height in cm}}{\text{creatinine (mg/dl)}}$$

Formula depends on:

- Height, muscle mass.
- Method of estimation of creatinine.
[Jaffei reaction].

S. creatinine is accurately measured by

- Enzyme assay / HPLC.

Value of k in Schwartz formula:

Low birth weight infant — 0.33

Normal infant 0-18 months — 0.45

Girls 2-16 yrs — 0.55

Boys 2-13 yrs — 0.55

Boys 13-16 yrs — 0.70

Schwartz method is independent of — Renal ~~function~~ function.

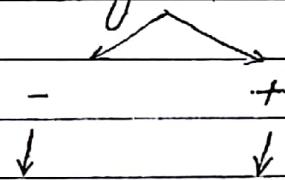
HEMATURIA:

Red urine:

- Beet root
- Phenolphthalein
- Rifaupicin

Q. Syms old, Red urine. Urine 1-2 RBCs/hpf —
Intravascular hemolysis.

If Red urine → Look for RBCs.



Hemoglobinuria Hematuria
Myoglobinuria.

Hematuria:

- Gross

- Microscope → > 5 RBCs/hpf on centrifuged urine



Cause —	Glomerular	Extrā glomerular
	- IgA ; MPGN; PSGN.	- Cystitis; Stones;

Idiopathic hypercalciuria
↓

24 hr urine $\text{Ca}^{2+} > 400 \text{ mg/kg}$.

Dysmorphic RBC ++

Colour Color Bright Red.

Pain Painless Painful.

Protein in urine Proteinuria

Recurrent Gross Hematuria:

- IgA
- MPGN
- Idiopathic hypercalciuria.

PSGN doesn't reoccur.

Alport's Syndrome:

Triad:

Eye: Ant. Leucocystosis

Ear: SNHL

Kidney: 75% Boys ESRD before < 30 yrs.

- 80% X linked > AR 15% > AD 5%
- Collagen IV → α -5 domain (in GB memb)

Good pasture Syndrome — α 3 domain of collagen IV abnormality.

INV: Electron Microscopy.

↳ Basement membr' is irregular.

↳ Splitting of the lamina densa

↳ Lamellation

↳ Striated GBM

↳ Basket weave appearance (classical)

Slit lamp exam → Keratoconus

Leuconicus

Acute Kidney Injury (AKI):

Best Biomarker → Urine NGAL

[Neutrophilic gelatinase associated lipocalcin].

② Urine IL-18

③ Urine KIM-1 (Kidney injury molecule)

④ Urine L-FABP (Fatty acid binding protein).

⑤ Serum cystatin-C

Types:

- Prerenal

- Renal

- Post renal

PRERENAL - Hypotension, Hypoxia

Newborns; hypovolemia, burns, diarrhea.

RENAL - ATN > HUS

POST RENAL - Obstruction.

Indices	Pre renal	-Renal
Urinary Sodium (meq/L)	< 20	> 40
Urine Osmolality	> 500	< 300
B-urea/ Creatinine ratio	> 20 : 1	< 20 : 1
Fractional excretion of Na%	< 1	> 1
$= \frac{\text{Urine Na}}{\text{Serum Na}} \times \frac{\text{Serum Cr}}{\text{Urine Cr}}$		

Cause of AKI:

M/c/c in children/Adults

Prerenal → ATN

↳ Hypovolaemia

Hypotension

Drugs ↳ Exo ↳ Toxins

Sepsis ↳

Hemoglobinuria ↳ Endo ↳

Myoglobinuria

Hemolytic Uremic Syndrome (HUS):

- Microangiopathic hemolytic anemia
- Thrombocytopenia
- AKI

• 90% follows diarrhea.

• Developed : E. coli O157:H7.

• Developing country : Shigella dysenteriae type I.

• Germany June 2011 : E. coli O104:H4

SHiga toxin → Cause Endothelial injury



TMA (Thrombotic microangiopathy).

On PBS → Schistocytes are classic of HUS

Rx : ECULIZUMAB [DOC] → for PNH

↳ drug against Cs.

If not available → Plasmapheresis.

Complications :

→ Insensible losses - 400 ml/m^2

- Fluid overload → Rx → Fluid restriction
- Hyperkalemia → cause arrhythmia / Sudden death.
- Dilutional hypo Na. Rx - Restrict fluid
- Dilutional anaemia Rx - PRBCs if Hb < 6
- Metabolic acidosis.
- Hyper PO₄ → Hypo Ca
- HTN.

Hyperkalemia :

Rx - Glucose & insulin intravenously



→ in 10-15 min.

- Ca²⁺ stabilises cardiac membrane potential.

↳ only given in ECG changes in Hyperkalemia.



Calcium Gluconate i.v.

~~Mechanism~~ : Hyperkalemia Rx :

Transcellular shift into cell.

- Insulin & Dextrose.

- Nebulised salbutamol.

Cardioprotective → i.v. Calcium Gluconate.

↑ delivery Na to distal

• Furosemide

• i.v. NaHCO₃

Dialysis . . .

K-binders \rightarrow Acetyle polystyrene.

CKD (Chronic kidney disease):

Causes in children -

- < 5 yrs - Hypoplasia
Dysplasia
Posterior urethral valves (Boys)
- > 5 yrs - Acquired
↳ GN/HUS.

Features:

A = Azotemia

Acidosis (metabolic)

Anemia (Normocytic Normochromic)

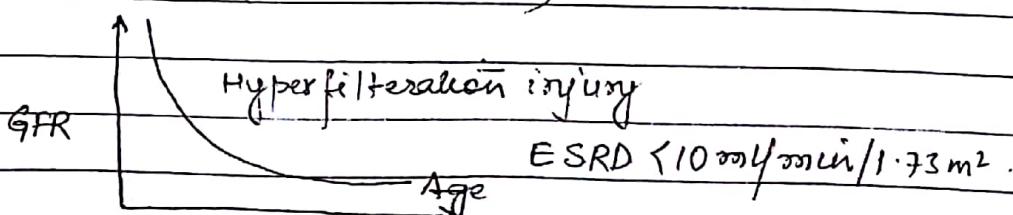
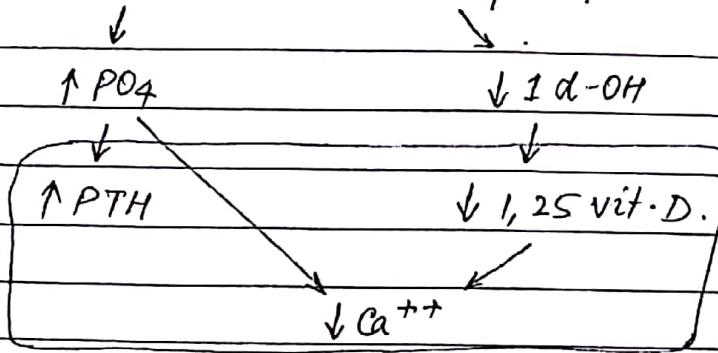
↳ Rx - S/C rh EPO (Erythropoietin).

B = Bone ds

C = Cardiovascular complication \uparrow

G = Growth failure \leftarrow Multifactorial causes.

When GFR \downarrow - 40-60 ml/min/1.73m²



Nephrotic Syndrome:

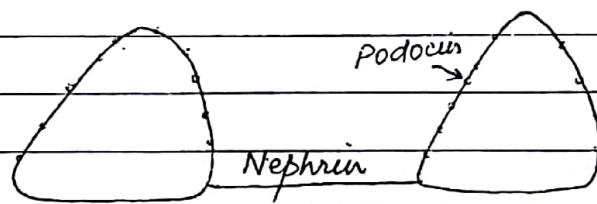
- Proteinuria $> 40 \text{ mg/m^2/hr}$ or $> 2 \text{ g/24 hr}$
- Hypoproteinemia (Hypoalbuminemia)
- Hyperlipidemia (Cholesterol $> 200 \text{ mg/dL}$)
- Edema: S. Albumin $< 2.5 \text{ g/dL}$

On light microscopy → Minimal changes.

On Electron " → Effacement of foot processes of podocytes.

DOC: Prednisolone

Cause of Edema in NS → Na^+ & H_2O reabsorption.



Gene	Protein	Disease
NPHS1 (Chr 19)	Nephrin	Finnish congenital nephrotic (< 3 months)
		Rx: Nephrectomy.
NPHS2 (Chr 1)	Podocin	Steroid resistant FSGS Rx: Catecholuria inhibitors ↳ Tacrolimus > Cyclosporine

Steroid toxic; Steroid dependent

- Cushingoid
- HTN
- Post- subcapular cataract
- Impaired glucose tolerance
- Short

Rx - Oral cyclophosphamide for 12 weeks.



UTI (Urinary Tract Infection):

Definition: Symptoms + Urine culture $> 10^5$ CFU/ml

- Symptom - Fever
- E. coli
- Females [Boys < 1 yr]
- Ascending [< 1 yr \rightarrow Hematogenous]

Best urine specimen - Suprapubic aspiration.

- Asymptomatic bacteriuria \rightarrow Shouldn't be treated.
- M/c/c of UTI in children \rightarrow VUR
IOC for VUR \rightarrow MCU

VUR:

- Polar scanning \rightarrow DMSA nuclear scan
- Function \rightarrow MAG3 / DTPA nuclear scan.
- Prophylaxis \rightarrow Antibiotic of choice

\downarrow
Cotrimoxazole.

Sx \rightarrow Reimplantation of both ureter.

\hookrightarrow Indication:

- Breakthrough UTIs
- Deterioration of Renal function.

GLOMERULONEPHRITIS:

- Hematuria
- Edema
- HTN

M/C/C of GN → Post streptococcal GN.

Acute post streptococcal GN:

- Follows infection of throat (serotype 12) / skin (serotype 49) of nephrogenic strains of β -hemolytic streptococci.
- Age → 5-12 yrs
- Acute phase resolves in 4-6 wks but urine normalises in 1 yr.

4-6 wks

Edema

X

Cala Gross hematuria

X

HTN

X

C₃

6-8 wks
normalise.

In urine - Shows microscopic hematuria

6-12 months to resolve.

Sore throat → Δ by ASO titre (1-2 wks)

Pyoderma → Δ by anti-DNAase B (4-6 wks)

95% PSGN resolves

5% PSGN → CKD

Kidney Bx → Endothelial & mesangial cell proliferation & obliteration of capillary lumen.

- Neutrophil infiltration

Immunofluorescence - Granular deposits of IgG & C3
 "STARRY SKY"

Good pasture Syndrome has linear deposits of IgG & C3.

Subepithelial humps are a sign of PSGN.

Q Persistently + C3 found in all except:

- a) Post streptococcal GN (normalise after 6-8 weeks)
- b) Mesangio capillary GN
- c) Cryoglobulinemia
- d) SLE
- e) IE
- f) Shunt nephritis
- g) Factor H-mutation → HUS.